Intramedullary schwannomas (IMS) represent exceptional rare pathologies. They commonly present as solitary lesions; only five cases of multiple IMS have been described so far. Here, we report the sixth case of a woman with multiple IMS. Additionally, we performed the first complete systematic review of the literature for all cases reporting IMS. We performed a systematic review of the literature in PubMed, EMBASE and Cochrane Central Register of Controlled (CENTRAL) to retrieve all relevant studies and case reports on IMS. In a second step, we analysed all reported studies with respect to additional cases, which were not identified through the database search. Studies published in other languages than English were included. One hundred nineteen studies including 165 reported cases were included. In only five cases, the patients harboured more than one IMS. Gender ratio showed a ratio of nearly 3:2 (male:female); mean age of disease presentation was 40.2 years; 11 patients suffered from neurofibromatosis (NF) type 1 or 2 (6.6%). IMS are rare. Our first systematic review on this pathology revealed 166 cases, including the here reported case of multiple IMS. Our review offers a basis for further investigation on this disease.

Keywords Schwannoma · Spinal tumour · Intramedullary tumour · Review of the literature

Introduction

Within the group of central nervous system tumours, spinal tumours represent a minor fraction of 15% of all cases [1]. Spinal schwannomas represent about 10% of all spinal tumours [1]. Schwannomas occur most frequently within the intradural-extradural compartment [1]. The intramedullary location of schwannomas is a rare condition (0.3–1.5%) [2–4]. Furthermore, they commonly present as solitary lesions. To date, only five cases of multiple intramedullary schwannomas (IMS) have been described [5–9].

Here, we report a 6th case of a female patient with histologically proven IMS of the cervical spinal cord and an additional small lumbar localized lesion. Additionally, we performed the first complete systematic review of the literature searching PubMed, EMBASE and Cochrane Central Register of Controlled Trials (CENTRAL) for all cases reporting IMS.

Case report

A 53-year-old woman presented with a 4-month history of progressive sensory deficits of the upper and lower limbs, without any further neurological symptoms. There were no neurofibromatosis (NF) stigmas and no history of genetic disorders or spinal injury.

Clinical presentation

Neurological examination revealed hypaesthesia of the first three fingers of the right hand, the right lateral lower leg and the right lateral foot edge. There was no paresis of the upper and lower limbs; the muscular tension was normal. The muscle stretch reflexes were normal and symmetrical. No pyramidal tract signs were present, nor spinal ataxia. The patient was defined as grade I according to the modified McCormick scale [10, 11].
Imaging findings and additional diagnostics

Magnetic resonance imaging (MRI) of the neurocranium and the cervical spine revealed a 9.3 × 19 mm intramedullary lesion at the level of C2/3, which was isointense on T1-weighted and had both hypo- and hyperintense components on T2-weighted images. The lesion showed intense heterogeneous contrast enhancement and caused a massive perilesional spinal cord edema extending from the medulla oblongata to the level of C6 (Fig. 1).

Combining the MRI findings and the neurological examination, we considered a preliminary diagnosis of intramedullary ependymoma. As a consequence, further investigations including a holospinal MRI and a lumbar puncture were carried out to examine the possible presence of drop metastasis. The holospinal MRI revealed a second small (3.4 × 4 mm) lesion at the level of L2/3. The lesion was isointense on T1-weighted and hypointense in T2-weighted images with homogeneous contrast enhancement (Fig. 1). Cerebrospinal fluid examination showed no evidence of atypical, potentially malignant cells.

Operative findings and histopathology

The patient underwent uneventful microsurgical tumour resection through a posterior cervical approach and midline myelotomy with subsequent C2–C3 laminoplasty. Intraoperatively, the tumour appeared as a solid, yellowish mass comparable with a schwannoma. Complete tumour resection was achieved via meticulous microsurgical technique and ultrasonic aspiration. Intraoperative monitoring (somatosensory-evoked potentials) remained stable during the entire surgical procedure.

Microscopic examination of tissue samples obtained during surgery showed spindle-shaped cells, arranged in a typical fascicular pattern. Old haemorrhages were frequently seen. Immunohistochemistry revealed a strong homogeneous reaction for S-100 protein but was negative for epithelial membrane antigen. The proliferation rate (Ki-67 staining) was low (Fig. 2). Altogether, these findings were consistent with a histopathological diagnosis of a schwannoma.

Postoperative recovery

Immediately after the surgery, the sensory and motor functions of the patient were intact. During the inpatient stay, the patient had a veritable postoperative course; the sensory impairments remained unchanged. Postoperative MRI of the cervical spine confirmed complete removal of the intramedullary lesion.

Fig. 1 a–c Preoperative MRI of the cervical spine in sagittal (a, b) and transverse (c) slides. T2-weighted images show a hypo-and hyperintense intramedullary lesion at the level of C2/3 (a). T1-weighted images show a heterogeneous gadolinium-enhanced tumour in the sagittal (b) and transverse (c) slides. d–f Preoperative MRI of the lumbar spine in sagittal (d, e) and transverse (f) slides. T2-weighted images show a hypointense lesion at the level of L2/3 (d). T1-weighted images show a homogenous gadolinium-enhanced tumour in the sagittal (e) and transverse (f) slides. g–i Postoperative MRI of the cervical spine in sagittal (g, h) and transverse (i) slides confirming the complete tumour resection.
Interestingly, the massive spinal cord edema decreased almost completely within 10 days after surgery (Fig. 1). The patient was discharged to medical rehabilitation. Follow-up examination 4 months after surgery revealed favourable, unchanged neurological condition (modified McCormick scale: grade I).

Material and methods
For this study, no experiments on human subjects or animals have been carried out. We performed a systematic review of the literature in PubMed, EMBASE and CENTRAL up to January 1, 2020, to retrieve all relevant studies and case reports on IMS. We used the keywords “intramedullary simultaneous with “schwannoma OR neurinoma”. Selection criteria were the following: (1) at least one histological proven IMS reported, (2) available clinical information of the patient and (3) peer reviewed publication in a journal or book chapter. Studies published in other languages than English were included in order to receive a complete review of all reported cases. Melanotic IMS were excluded because of their reclassification as a distinct entity in 2016 [12]. In a second step, for complete identification, all reported studies on IMS have been analysed regarding additional cases of IMS. Each case which was mentioned in these articles was analysed with respect to our inclusion criteria. If not already found via keyword search, the case was added to our systematic review (Fig. 3).

Results
One hundred nineteen studies including 165 reported cases met our inclusion criteria. In only five cases, the patients harboured more than one IMS. Gender ratio was nearly 3:2 (male: female; 55.4% male; 39.2% female); mean age of disease presentation was 40.2 years (range 1 day–78 years); eleven patients suffered from NF (6.6%). A closer analysis of patients suffering from NF revealed that one patient had NF type 1, eight patients had NF type 2 and in two cases no information on the NF type was available. Most IMS were located in the cervical (45.8%) and thoracic (37.3%) spine; a smaller number was located in the cervicothoracic (6.2%), thoracolumbar (5.6%) and lumbar (2.3%) spine (Table 1).
We reviewed the included cases with respect to preoperative neurological status, the postoperative outcome and the follow-up, including tumour recurrence. In addition, we calculated the modified McCormick scale to determine the neurological status preoperatively and postoperatively. The analysis of preoperative neurological symptoms showed that sensory disturbance appeared in 67%, motor deficits in 68% and dysfunction of the autonomic nervous system, such as sphincter dysfunction, in 26% of the cases. The main duration of symptoms was 29 months. The preoperative neurological status according to the modified McCormick scale showed the following distribution: grade I (6%), grade II (27%), grade III (21%), grade IV (12%) and grade V (4%); in 30% of the cases, the preoperative modified McCormick scale was not determinable (Table 1).

Our review showed that 161 of 165 patients underwent surgery; in four cases, the diagnosis of IMS was made postmortem by autopsy. The analysis of the postoperative recovery revealed that complete recovery was achieved in 23%, symptom improvement in 51% and stable neurological condition in 4% of the cases. The neurological symptoms worsened in only 4% of cases and in another 4% the patient died after surgery. Information on the postoperative recovery was missing in 14% of the cases. The postoperative neurological status
| Case No. | Reference          | Patient Characteristics | Recovery McCormick scale* | Follow-up | Tumour recurrence |
|---------|--------------------|-------------------------|---------------------------|-----------|------------------|
|         |                    | Age | Sex | NF  | Vertebra | Sensory system | Motor system | Autonomic nervous system | Duration (months) | Follow-up | Months | McCormick scale* | Tumour recurrence |
| 1       | Penfield, 1932 [13]| 12  | M   | No  | C5  | Yes      | Yes         | No            | 96          | Yes       | n.a.   | n.a.   | n.a.   | n.a.   |
| 2       | Rasmussen et al., 1940 [14]| 12  | M   | No  | C4–7| n.a.    | n.a.        | n.a.        | 48          | Yes       | –      | n.a.   | n.a.   | n.a.   |
| 3       | Roka, 1951 [15]    | 30  | M   | No  | Cerv.| n.a.    | n.a.        | n.a.        | 36          | Yes       | n.a.   | n.a.   | n.a.   | n.a.   |
| 4       | Rose, 1954 [16]    | 61  | M   | NF  | 1   | C5      | n.a.        | n.a.        | 96          | Yes       | n.a.   | n.a.   | n.a.   | n.a.   |
| 5       | Riggs/Clary, 1957 [17]| 60  | M   | No  | C4/5| Yes     | Yes         | Yes         | n.a.        | Yes       | –      | IV     | IV     | 24     | n.a.   |
| 6       | Ramamurthi et al., 1958 [18]| 35  | M   | No  | T2   | Yes     | Yes         | Yes         | 9           | Yes       | +      | V      | III    | 48     | III    |
| 7       | Scott/Bentz, 1962 [19]| 46  | F   | No  | T3   | Yes     | Yes         | No           | 144         | Yes       | o      | V      | V      | n.a.   | n.a.   |
| 8       | McCormick et al., 1964 [20]| 62  | M   | No  | L2   | No      | No          | No           | n.a.        | No        | n.a.   | n.a.   | n.a.   | n.a.   |
| 9       | Sloof, 1964 [9]    | 62  | F   | No  | Cerv.| Yes     | No          | No           | n.a.        | No (Autopsy) | n.a.   | n.a.   | n.a.   | n.a.   |
| 10      | Mason/Keigher, 1968 [21]| 37  | M   | No  | T8–10| Yes     | Yes         | No           | 3           | Yes       | +      | III    | III    | 6      | II     |
| 11      | Chigasaki/Pennybacker, 1968 [22]| 75  | F   | No  | T3   | Yes     | Yes         | No           | 7           | Yes       | –      | V      | V      | 6      | n.a.   |
| 12      | Van Duinen, 1971 [23]| 24  | F   | No  | C3   | Yes     | Yes         | Yes          | 48          | Yes       | +      | III    | IV     | 3      | II     |
| 13      | Fabres et al., 1972 [24]| 26  | M   | No  | T2/3| Yes     | Yes         | No           | 13          | Yes       | +      | IV     | IV     | n.a.   | n.a.   |
| 14      | Cambier et al., 1974 [25]| 60  | M   | No  | C2–4| Yes     | Yes         | No           | 6           | Yes       | –      | III    | IV     | 17     | IV     |
| 15      | Wood et al., 1975 [26]| 48  | M   | No  | C3   | Yes     | Yes         | No           | 3           | Yes       | –      | IV     | IV     | 0      | n.a.   |
| 16      | Schmitt, 1975 [27]| 68  | M   | No  | L1   | Yes     | Yes         | No           | n.a.        | No (Autopsy) | n.a.   | n.a.   | n.a.   | n.a.   |
| 17      | Isu et al., 1976 [28]| 30  | F   | No  | Cl   | Yes     | Yes         | No           | 6           | Yes       | n.a.   | III    | n.a.   | n.a.   | n.a.   |
| 18      | Kumar/Galati, 1977 [29]| 24  | F   | NF  | Cerv.| Yes     | Yes         | No           | 12          | Yes       | o      | V      | V      | n.a.   | n.a.   |
| 19      | Vailati et al., 1979 [30]| 40  | F   | No  | T8/9| No      | Yes         | No           | 12          | Yes       | +      | IV     | IV     | 6      | II     |
| 20      | Gegalian, 1979 [31]| 37  | F   | No  | T10/11| Yes    | Yes         | No           | n.a.        | Yes       | +      | IV     | IV     | 120    | II     |
| 21      | Pandatscher et al., 1979 [8]| 41  | M   | No  | T2–8| Yes     | Yes         | Yes          | 6           | Yes       | –      | IV     | III    | n.a.   | n.a.   |
| 22      | Shalit/Sandbank, 1981 [32]| 21  | F   | No  | C2-T2| Yes     | Yes         | No           | 6           | Yes       | +      | IV     | III    | 18     | II     |
| 23      | Guidetti, 1967 [33], Cantore et al., 1982 [34]| 54  | F   | No  | C3–5| Yes     | Yes         | No           | 24          | Yes       | +      | II     | I      | n.a.   | n.a.   |
| 24      |                     | 57  | M   | No  | T12–L1| No      | No          | No           | n.a.        | Yes       | +      | I      | I      | n.a.   | n.a.   |
| Case No. | Reference | Patient Age | Sex | NF | Vertebra | Sensory system | Motor system | Autonomic nervous system | Duration (months) | McCormick scale* | Follow-up | Recovery | McCormick scale* | Tumour recurrence |
|----------|-----------|-------------|-----|----|----------|---------------|-------------|-------------------------|-----------------|----------------|-----------|----------|----------------|------------------|
| 25       | Lesoin et al., 1983 [35] | 45 F | No | C3–7 | No | No | No | 6 | Yes | + | n.a. | II | n.a. | n.a. | n.a. |
| 26       | Rout et al., 1983 [36] | 28 M | No | L1 | No | Yes | Yes | 50 | Yes | + | n.a. | III | 11 | II | No |
| 27       | Kang/Song, 1983 [37] | 50 F | No | C3–5 | Yes | Yes | Yes | 60 | Yes | + | III | 12 | II | No | |
| 28       | Bouchez et al., 1984 [38] | 47 M | No | C3–6 | Yes | Yes | No | 12 | Yes | + | IV | III | 6 | II | No |
| 29       | Drapkin et al., 1985 [39] | 34 M | No | C2–7 | Yes | Yes | No | 12 | Yes | – | II | II | 60 | IV | No |
| 30       | Rout et al., 1983 [36] | 30 F | No | C3–5 | Yes | Yes | No | 46 | Yes | + | II | I | 20 | I | No |
| 31       | Lesoin et al., 1986 [40] | 75 M | No | T3–6 | Yes | Yes | Yes | 60 | Yes | + | IV | III | 6 | III | No |
| 32       | Maniki et al., 1986 [41] | 42 F | No | T7/8 | Yes | Yes | No | n.a. | Yes | + | n.a. | n.a. | n.a. | n.a. | n.a. |
| 33       | Ross et al., 1986 [4] | 67 F | No | C2–T1 | Yes | Yes | Yes | 48 | Yes | + | II | I | 6 | I | No |
| 34       | Char/Cross, 1987 [42] | 36 M | No | C4/5 | Yes | Yes | No | 4 | Yes | + | II | I | n.a. | n.a. | n.a. |
| 35       | Garen et al., 1988 [43] | 54 M | No | T3/4 | Yes | Yes | Yes | 1 | Yes | – | II | I | 0 | n.a. | n.a. |
| 36       | Hida et al., 1988 [44] | 30 F | No | C3–6 | Yes | Yes | Yes | 24 | Yes | + | II | II | n.a. | n.a. | n.a. |
| 37       | Okuda et al., 1988 [45] | 72 F | No | T8/9 | Yes | Yes | Yes | 132 | Yes | + | IV | III | 6 | III | No |
| 38       | Gorman et al., 1989 [46] | 23 M | No | Med.–C7 | Yes | Yes | No | n.a. | Yes | + | IV | III | 6 | III | No |
| 39       | Sharma et al., 1989 [47] | 15 F | No | C5/6 | Yes | Yes | No | 8 | Yes | + | II | III | 5 | II | No |
| 40       | Herregodts et al., 1991 [50] | 10 M | No | C5 | Yes | Yes | Yes | 12 | Yes | + | IV | III | 6 | II | No |
| 41       | Meisel et al., 1990 [48] | 36 M | No | T9/10 | Yes | Yes | Yes | 36 | Yes | ++ | III | II | 2 | I | No |
| 42       | Li/Holtas, 1991 [49] | 67 F | n.a. | C2 | n.a. | n.a. | n.a. | n.a. | Yes | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 43       | Jacquet et al., 1992 [51] | 49 F | No | T2 | No | Yes | Yes | 60 | Yes | + | III | III | 2 | II | No |
| 44       | Morimoto et al., 1992 [52] | 44 M | No | T12 – L1 | No | No | No | 5 | Yes | ++ | I | I | n.a. | n.a. | n.a. |
| 45       | Benini et al., 1993 [53] | 42 M | No | T7–9 | No | Yes | No | 13 | Yes | + | II | I | n.a. | n.a. | n.a. |
| 46       | Sereci et al., 1993 [54] | 40 M | No | T7–9 | Yes | Yes | Yes | 36 | Yes | + | III | IV | 5 | II | No |
| 47       | Radhakrishnan et al., 1993 [55] | 43 M | No | C5/6 | Yes | No | Yes | 60 | Yes | – | I | IV | 12 | IV | No |
| 48       | Sekerci et al., 1993 [54] | 45 F | No | T1–3 | Yes | Yes | No | 4 | Yes | o | II | IV | 6 | II | No |
| 49       | Nicoletti et al., 1994 [56] | 50 F | No | C2–5 | Yes | Yes | No | 60 | Yes | + | IV | II | 12 | II | No |
| 50       | Duong et al., 1995 [57] | 55 M | No | C4–6 | Yes | Yes | No | 12 | Yes | + | II | I | 3 | II | No |
| 51       | Melancia et al., 1996 [58] | 47 F | No | C3–5 | No | Yes | No | 6 | Yes | + | V | III | 12 | n.a. | No |
| 52       | Tu et al., 1995 [57] | 52 F | No | T5–7 | No | Yes | No | 18 | Yes | ++ | II | I | 60 | n.a. | Yes |
| 53       | Melancia et al., 1996 [58] | 53 F | No | T11–L2 | No | Yes | No | 24 | Yes | – | II | V | 36 | V | Yes |
| 54       | Lee et al., 1996 [2] | 31 F | No | C5–T3 | n.a. | n.a. | n.a. | n.a. | Yes | n.a. | n.a. | n.a. | 12 | II | n.a. |
Table 1 (continued)

| Case No. | Reference                  | Patient Age | Sex | NF | Vertebra | Sensory system | Motor system | Autonomic nervous system | Duration (months) | McCormick scale* Prior OP | McCormick scale* Post OP | Follow-up | Tumour recurrence |
|----------|----------------------------|-------------|-----|----|----------|----------------|--------------|---------------------------|------------------|-----------------------------|---------------------------|-----------|-------------------|
| 56       | Bhayani/Goel, 1996 [6]     | 15 M        | NF  | C4/5 | Yes     | No             | Yes          | No                        | 18               | Yes +                        | III II                    | 2 I No       | No                |
| 57       | Botelho et al., 1996 [59]  | 52 F        | No  | C4–6 | Yes     | Yes            | No           | No                        | 48               | Yes +                        | III n.a.                  | 12 II No     | No                |
| 58       | Innocenzi et al., 1996 [60]| 44 M        | No  | C1–3 | No      | Yes            | No           | No                        | 18               | Yes ++                       | II II                     | 24 I No      | No                |
| 59       | Bekar et al., 1997 [61]    | 40 M        | No  | C2–T1| Yes     | Yes            | Yes          | Yes                       | 60               | Yes n.a.                     | II III                    | 12 III No    | No                |
| 60       | Böskonakli et al., 1997 [62]| 42 F      | No  | T8   | Yes     | Yes            | No           | Yes                       | 12               | Yes +                        | III II                    | 12 II No     | No                |
| 61       | Chitojuku et al., 1998 [63]| 26 M        | NF  | T4/5 | Yes     | Yes            | No           | n.a.                      | 120              | Yes ++                       | III III                   | n.a. n.a.    | No                |
| 62       | Kotil et al., 1998 [64]    | 20 F        | NF  | T10/11| n.a.    | n.a.           | n.a.         | Yes n.a.                   | 12               | III n.a.                     | n.a. n.a.                 | 0 n.a. n.a.  | No                |
| 63       | Hejazi/Hassler, 1998 [65]  | 65 M        | No  | T12–L1| Yes     | Yes            | Yes          | No                        | 120              | Yes ++                       | n.a. n.a.                  | 0 n.a. n.a.  | No                |
| 64       | Binatlı et al., 1999 [66]  | 9 M         | No  | C6–T1| Yes     | Yes            | Yes          | Yes                       | 4                | Yes ++                       | II I                     | 3 I No       | No                |
| 65       | Arellanes-Chávez et al., 2000 [67]| 18 M      | No  | C2–5 | No      | Yes            | No           | 36                        | Yes              | +                            | II II                    | n.a. n.a.    | No                |
| 66       | Riffaud et al., 2000 [3]   | 25 M        | No  | C1/2 | Yes     | Yes            | No           | 12                        | Yes              | +                            | III III                   | 12 II No     | No                |
| 67       | Ogumbgo et al., 2000 [68]  | 24 M        | No  | C4–7 | Yes     | Yes            | No           | 36                        | Yes              | +                            | III n.a.                  | 18 II No     | No                |
| 68       | Kodama et al., 2000 [69]   | 37 F        | No  | C3–5 | Yes     | Yes            | No           | 108                       | Yes              | +                            | n.a. n.a.                  | n.a. n.a.    | No                |
| 69       | 17 F                       | No  | C1   | Yes     | Yes            | Yes          | 12                        | Yes              | +                            | n.a. n.a.                  | n.a. n.a.    | No                |
| 70       | Patronas et al., 2001 [70] | 26 n.a.     | NF  | 2 n.a.| n.a.    | n.a.           | n.a.         | n.a.                      | Yes              | n.a.                        | n.a. n.a.                  | n.a. n.a.    | n.a. n.a. |
| 71       | Kono et al., 2001 [71]     | 59 M        | No  | T2   | Yes     | Yes            | No           | 6                         | Yes              | +                            | n.a. n.a.                  | n.a. n.a.    | n.a. n.a. |
| 72       | Maira et al., 2001 [72]    | 69 M        | No  | C2   | Yes     | Yes            | Yes          | n.a.                      | Yes              | ++                           | III I                     | 36 n.a. No   | No                |
| 73       | Sasaki et al., 2002 [73]   | 53 M        | NF  | C5/6 | Yes     | Yes            | No           | n.a.                      | Yes              | +                            | II II                    | n.a. n.a.    | No                |
| 74       | Darwish et al., 2002 [74]  | 68 F        | No  | C3/4 | Yes     | Yes            | No           | 108                       | Yes              | o                            | II II                     | n.a. n.a.    | No                |
| 75       | Brown et al., 2002 [75]    | 51 F        | No  | T3–8 | Yes     | Yes            | No           | 24                        | Yes              | +                            | III IV                    | 6 III No     | No                |
| 76       | O’Brien et al., 2003 [76]  | 48 M        | No  | T11–L1| Yes     | Yes            | No           | 6                         | Yes              | ++                           | I I                     | 6 I No       | No                |
| 77       | Colosimo et al., 2003 [77] | 59 M        | No  | C2   | Yes     | Yes            | No           | 12                        | Yes              | +                            | n.a. n.a.                  | 48 I No      | No                |
| 78       | 47 F                       | No  | T8   | No      | Yes            | No           | 12                        | Yes              | +                            | n.a. III                   | 36 II No     | No                |
| 79       | Panagiotopoulos et al., 2004 [78]| 71 M      | No  | T6   | Yes     | Yes            | No           | 12                        | Yes              | ++                           | IV II                    | 36 I No      | No                |
| 80       | 51 M                       | No  | T9/10| Yes     | Yes            | No           | 3                         | Yes              | +                            | IV II                    | n.a. n.a.    | No                |
| 81       | Siddiqui/Shah, 2004 [79]   | 13 F        | NF  | Med.–C3| Yes    | Yes            | No           | 6                         | Yes              | +                            | III n.a.                  | 3 II No      | No                |
| 82       | Conti et al., 2004 [80]    | 28 F        | NF  | C1   | Yes     | Yes            | n.a.         | Yes                       | n.a.              | IV n.a.                     | n.a. n.a.                  | n.a. n.a.    | No                |
| 83       | 31 F                       | No  | C4–6 | n.a.    | n.a.      | n.a.         | 72                        | Yes              | +                            | n.a. n.a.                  | n.a. n.a.    | Yes               |
| 84       | 44 M                       | No  | T10  | n.a.    | n.a.      | n.a.         | 36                        | Yes              | +                            | n.a. n.a.                  | n.a. n.a.    | No                |
| Case No. | Reference | Patient Age | Sex | NF | Vertebra | Sensory system | Motor system | Autonomic nervous system | Duration (months) | Recovery McCormick scale* | Follow-up Months | McCormick scale* | Tumour recurrence |
|----------|-----------|-------------|-----|----|----------|---------------|--------------|------------------------|------------------|-----------------------------|----------------|----------------|-------------------|
| 85       | Chavez-Lopez et al., 2004 [81] | 40 | M | No | C4–6 | Yes | Yes | No | 24 | Yes | II | I | n.a. | n.a. | No |
| 86       | El Malki et al., 2005 [82] | 40 | F | No | C1–6 | Yes | Yes | No | 84 | Yes | + | I | I | n.a. | n.a. | 6 | n.a. | No |
| 87       | Anato et al., 2005 [83] | 38 | F | No | C4 | Yes | No | No | 1 | Yes | + | I | I | n.a. | n.a. | 36 | I | No |
| 88       | Matsuyama et al., 2009 [84], Kim et al., 2005 [85] | 72 | F | No | T8/9 | Yes | No | No | 10 | Yes | + | II | II | n.a. | n.a. | n.a. | No |
| 89       | Kyoshima et al., 2005 [86] | 54 | M | No | T9/10 | Yes | Yes | Yes | 48 | Yes | + | II | III | 60 | II | No |
| 90       | Shenoy/Raja, 2005 [87] | 29 | M | No | C4–7 | Yes | Yes | Yes | 36 | Yes | + | n.a. | n.a. | n.a. | n.a. | No |
| 91       | Kahilogullari et al., 2005 [88] | 38 | F | No | T12–L2 | Yes | No | No | 7 | Yes | + | I | I | n.a. | n.a. | n.a. | No |
| 92       | Ho et al., 2006 [89] | 45 | M | No | C5/6 | No | No | No | n.a. | Yes | + | I | I | 4 | I | No |
| 93       | Mukerji et al., 2007 [90] | 8 | M | No | C5–7 | Yes | Yes | Yes | 6 | Yes | + | V | n.a. | 18 | I | No |
| 94       | Hida et al., 2008 [91] | 41 | M | No | C1/2 | Yes | Yes | Yes | 6 | Yes | + | n.a. | n.a. | n.a. | n.a. | No |
| 95       | 30 | M | No | C5–7 | Yes | Yes | No | n.a. | Yes | + | n.a. | n.a. | n.a. | n.a. | No |
| 96       | Kim et al., 2009 [92] | 11 | F | No | T5/6 | Yes | Yes | Yes | 9 | Yes | – | II | IV | 138 | III | No |
| 97       | Nicácio et al., 2009 [93] | 40 | M | No | C4–6 | Yes | Yes | Yes | 24 | Yes | + | III | III | 24 | III | No |
| 98       | Hayashi et al., 2009 [94] | 78 | F | No | T11–L1 | Yes | Yes | No | 240 | Yes | + | I | I | 10 | III | No |
| 99       | Ohtonari et al., 2009 [95] | 29 | M | No | T12–L1 | No | Yes | Yes | 8 | Yes | + | II | I | n.a. | n.a. | n.a. | n.a. | No |
| 100      | Adam et al., 2010 [96] | 21 | F | No | C2–5 | Yes | Yes | Yes | 18 | Yes | ++ | II | I | 12 | I | No |
| 101      | 46 | F | No | T2–6 | n.a. | n.a. | n.a. | 6 | Yes | + | III | n.a. | 48 | n.a. | No |
| 102      | Lyle et al., 2010 [97] | 0 | M | No | T2–Sacr. | Yes | Yes | n.a. | n.a. | Yes | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | No |
| 103      | Bernal-Garcia et al., 2010 [5] | 35 | F | No | T1–5 | Yes | Yes | Yes | 36 | Yes | + | III | n.a. | 204 | II | No |
| 104      | 18 | F | NF 2 | C5–7 | Yes | Yes | Yes | 24 | Yes | + | III | n.a. | n.a. | II | No |
| 105      | Teo et al., 2011 [98] | 44 | M | No | C5/6 | Yes | Yes | No | 24 | Yes | + | II | I | n.a. | n.a. | n.a. | No |
| 106      | Ryu et al., 2011 [99] | 68 | M | No | T6/7 | Yes | Yes | No | 17 | Yes | + | III | III | 1 | II | No |
| 107      | Vij et al., 2011 [100] | 25 | M | No | T10/11 | Yes | Yes | Yes | 36 | Yes | – | III | IV | n.a. | n.a. | n.a. | No |
| 108      | Das et al., 2012 [101] | 55 | M | No | C2/3 | No | No | No | n.a. | Yes | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | No |
| 109      | Li et al., 2013 [102] | 42 | M | No | T10/11 | Yes | Yes | Yes | 18 | Yes | + | IV | IV | 18 | I | No |
| 110      | Lee et al., 1999 [103], Lee et al., 2013 [104] | 39 | F | No | T1–4 | n.a. | n.a. | n.a. | n.a. | Yes | + | n.a. | n.a. | n.a. | n.a. | No |
| 111      | 41 | F | No | C5/6 | n.a. | n.a. | n.a. | n.a. | Yes | + | n.a. | n.a. | n.a. | n.a. | No |
| 112      | 49 | F | No | C5–7 | n.a. | n.a. | n.a. | n.a. | Yes | + | n.a. | n.a. | n.a. | n.a. | No |
| 113      | 46 | F | No | T1/2 | n.a. | n.a. | n.a. | n.a. | Yes | + | n.a. | n.a. | n.a. | n.a. | No |
| 114      | 19 | F | No | T6–8 | n.a. | n.a. | n.a. | n.a. | Yes | + | n.a. | n.a. | n.a. | n.a. | No |
| Case No. | Reference Patient | Localization | Symptoms | OP | Recovery McCormick scale* | Follow-up | Tumour recurrence |
|---------|-------------------|--------------|----------|----|--------------------------|-----------|------------------|
|          |                   | Age | Sex | NF | Vertebral | Sensory system | Motor system | Autonomic nervous system | Duration (months) | PréOP | Post OP | Months | McCormick scale* | Tumour recurrence |
| 115      |                   | 42  | M   | No | T7/8       | n.a.   | n.a.         | n.a.         | Yes                | +     | n.a. | n.a. | n.a. | n.a. | No          |
| 116      |                   | 60  | No  | T7–10 | n.a. | n.a. | n.a. | n.a. | Yes | + | n.a. | n.a. | n.a. | n.a. | No |
| 117      |                   | 44  | M   | No | T8/9      | n.a.   | n.a.         | n.a.         | Yes                | + | n.a. | n.a. | n.a. | n.a. | No |
| 118      |                   | 37  | F   | No | T9/10     | n.a.   | n.a.         | n.a.         | Yes                | + | n.a. | n.a. | n.a. | n.a. | No |
| 119      |                   | 78  | M   | No | T10/11    | n.a.   | n.a.         | n.a.         | Yes                | + | n.a. | n.a. | n.a. | n.a. | No |
| 120      | Eljebbouri et al., 2013 [105] | 10  | M   | No | T7–9      | Yes | Yes | Yes | 6 | Yes | + | III | n.a. | 18 | I | No |
| 121      | Wu et al., 2011 [106], Yang et al., 2014 [107] | 52  | M   | No | C6–T4    | No | Yes | Yes | 120 | Yes | o | III | III | 154 | III | No |
| 122      |                   | 41  | F   | No | C4–6      | No | Yes | No | 6 | Yes | ++ | II | II | 140 | I | No |
| 123      |                   | 39  | M   | No | C3–5      | Yes | No | No | 12 | Yes | ++ | I | I | 125 | I | No |
| 124      |                   | 35  | M   | No | C6        | Yes | No | No | 36 | Yes | ++ | I | II | 114 | I | No |
| 125      |                   | 46  | M   | No | T3–5      | Yes | Yes | No | 12 | Yes | + | III | III | 102 | II | No |
| 126      |                   | 61  | M   | No | C6/7      | Yes | No | No | 24 | Yes | ++ | II | I | 94 | I | No |
| 127      |                   | 42  | M   | No | T10–12    | Yes | No | No | 24 | Yes | ++ | III | II | 85 | I | No |
| 128      |                   | 31  | M   | No | C3/4      | Yes | No | No | 12 | Yes | ++ | II | I | 78 | I | No |
| 129      |                   | 56  | F   | No | C5/6      | Yes | Yes | No | 36 | Yes | ++ | II | III | 74 | I | No |
| 130      |                   | 60  | F   | No | T2/3      | Yes | No | No | 36 | Yes | ++ | II | I | 65 | I | No |
| 131      |                   | 48  | M   | No | T9/10     | Yes | Yes | Yes | 144 | Yes | + | III | IV | 58 | III | No |
| 132      |                   | 59  | M   | No | C1/2      | Yes | No | No | 36 | Yes | ++ | I | III | 54 | I | No |
| 133      |                   | 50  | F   | No | C5/T1     | Yes | No | No | 24 | Yes | ++ | II | III | 51 | I | No |
| 134      |                   | 57  | M   | No | C4–6      | No | No | No | 6 | Yes | ++ | II | II | 47 | I | No |
| 135      |                   | 44  | F   | No | C5–7      | Yes | No | No | 48 | Yes | ++ | II | II | 41 | I | No |
| 136      |                   | 44  | M   | No | T3        | Yes | Yes | No | 12 | Yes | ++ | II | II | 24 | I | No |
| 137      |                   | 40  | M   | No | C3        | Yes | No | No | 2 | Yes | ++ | II | II | 20 | I | No |
| 138      |                   | 34  | M   | No | T12       | No | Yes | No | 48 | Yes | ++ | II | II | 16 | I | No |
| 139      |                   | 17  | M   | No | T6–8      | Yes | No | No | 12 | Yes | ++ | II | III | 12 | I | No |
| 140      |                   | 38  | M   | No | T11       | Yes | No | Yes | 18 | Yes | ++ | III | II | 6 | I | No |
| 141      | Yang et al., 2015 [108] | 35  | M   | No | T11/12    | Yes | Yes | Yes | 24 | Yes | ++ | II | II | 3 | I | No |
| 142      | Gupta et al., 2015 [109] | 48  | M   | No | C3/4      | Yes | Yes | No | 5 | Yes | + | III | III | 12 | II | No |
| 143      | Jagannatha et al., 2016 [110] | 11  | M   | No | T11/12    | Yes | Yes | Yes | 12 | Yes | ++ | III | n.a. | 6 | n.a. | No |
| 144      | Sun et al., 2017 [111] | 24  | M   | n.a. | C1/2   | Yes | Yes | Yes | 6 | Yes | + | II | I | n.a. | n.a. | n.a. |
### Table 1 (continued)

| Case No. | Reference | Age | Sex | NF | Localization | Symptoms | OP | Recovery McCormick scale* | Follow-up | Tumour recurrence |
|----------|-----------|-----|-----|----|--------------|----------|----|----------------------------|-----------|------------------|
| 145      | Nayak et al., 2017 [112] | 28  | M   | No | T1–9 | Yes Yes Yes | 36 | Yes + | IV III n.a. n.a. n.a. |          |
| 146      | Gao et al., 2017 [113] | 34–59 | 6 M 2 F | No | T8/9 | n.a. n.a. n.a. | n.a. | Yes n.a. | n.a. n.a. n.a. n.a. n.a. |          |
| 147      | No | T9/10 | n.a. n.a. n.a. | n.a. | Yes n.a. | n.a. n.a. n.a. n.a. n.a. n.a. |          |
| 148      | No | T10 | n.a. n.a. n.a. | n.a. | Yes n.a. | n.a. n.a. n.a. n.a. n.a. n.a. |          |
| 149      | No | T4–6 | n.a. n.a. n.a. | n.a. | Yes n.a. | n.a. n.a. n.a. n.a. n.a. n.a. |          |
| 150      | No | T10/11 | n.a. n.a. n.a. | n.a. | Yes n.a. | n.a. n.a. n.a. n.a. n.a. n.a. |          |
| 151      | No | C6–T1 | n.a. n.a. n.a. | n.a. | Yes n.a. | n.a. n.a. n.a. n.a. n.a. n.a. |          |
| 152      | No | C5/6 | n.a. n.a. n.a. | n.a. | Yes n.a. | n.a. n.a. n.a. n.a. n.a. n.a. |          |
| 153      | No | C4–7 | n.a. n.a. n.a. | n.a. | Yes n.a. | n.a. n.a. n.a. n.a. n.a. n.a. |          |
| 154      | Karatay et al., 2017 [114] | 30  | F   | No | T12/L1 | No Yes No | 2 | Yes + | II n.a. n.a. n.a. n.a. |          |
| 155      | Li et al., 2017 [115] | 30  | M   | No | C3–5 | No No No | 1 No n.a. n.a. n.a. n.a. |          |
| 156      | Navarro Fernández et al., 2018 [116] | 19  | M   | No | C6–7 | Yes Yes No | 36 | Yes + | IV IV 1 III No |
| 157      | Landi et al., 2018 [117] | 8   | F   | No | T10/11 | Yes Yes No | 8 | Yes ++ | III II 84 I No |
| 158      | Singh et al., 2018 [118] | 27  | F   | No | T12–L2 | Yes Yes Yes | 12 | Yes + | III III 6 III No |
| 159      | Wang et al., 2018 [119] | 9   | M   | No | T8 | Yes Yes No | 6 | Yes ++ | II I 36 I No |
| 160      | Shi et al., 2019 [120] | 42  | F   | No | Cerv. | n.a. n.a. n.a. | n.a. | Yes n.a. | n.a. n.a. n.a. 36 n.a. n.a. |          |
| 161      | Dhake/Chatterjee, 2019 [121] | 10  | M   | No | T10–12 | Yes Yes Yes | 6 | Yes + | III n.a. 216 V Yes |
| 162      | 57  | F   | No | T9/10 | Yes Yes No | 24 | Yes + | IV n.a. 24 III Yes |
| 163      | Dai et al., 2019 [122] | 34  | M   | No | C3/4 | Yes Yes No | 24 | Yes ++ | I I 12 I No |
| 164      | Sekar et al., 2019 [123] | 37  | F   | No | C5–7 | Yes Yes Yes | 12 | Yes n.a. | II n.a. n.a. n.a. n.a. n.a. |          |
| 165      | Kelly et al., 2020 [124] | 43  | M   | No | C4-T2 | Yes Yes No | 18 | Yes + | V V 3 IV No |

*Modified McCormick scale

n.a. information not available

NF neurofibromatosis

OP operation
our own study is the largest review of cases on IMS. An
research of the databases. This series of 166 cases including
case series revealed cases, which were missed by keyword
tabases did not show all cases; further analysis of reported
Portuguese, German and Spanish were not included in previ-

The average duration of follow-up on a patient with IMS
was 34 months. Tumour recurrence was only observed in 4% of
the cases (Table 1).

Information on MRI images were available in only half of
the cases. In the available T_1-weighted images, most cases
showed an isointense (18.1%) or hypointense (16.9%) imaging
pattern; mixed (6.8%) and hyperintense (6.2%) patterns
were observed less frequently. T_2-weighted images showed in
23.2% a hyperintense, in 11.9% an isointense, in 8.5% a
mixed and in 7.9% a hypointense pattern. All cases showed
a gadolinium enhancement, which was homogenous in
32.8%, heterogenous in 18.6%, some cases showed only a
circular (5.6%) and 2 cases were reported to only show min-
imal gadolinium enhancement (1.1%). 17.5% of the IMS
showed a cystic component. Perifocal edema was observed
in 22% of the cases; 20.9% of cases were associated with syringomyelia (Table 2).

Discussion

To our knowledge, no complete review of all reported cases
has been performed thus far. Here, we attempted to gather all
reported cases since 1932. Interestingly, we found more cases
than previously described in other series [62, 80, 98]. Due to
the language barrier, reports in Japanese, Chinese, French,
Portuguese, German and Spanish were not included in previ-
ous reports. Additionally, keyword research in the known da-
tabases did not show all cases; further analysis of reported
case series revealed cases, which were missed by keyword
research of the databases. This series of 166 cases including
our own study is the largest review of cases on IMS. An
uncomplete review of this very rare pathology might consti-
tute a limitation, which impacts the estimated epidemiology.

IMS represent 0.3–1.5% of all spinal schwannomas [2–4].
Several studies described a gender distribution of 3:1
(male:female) [93, 107, 113]. Our results showed a higher rate
of female patients and thus a gender distribution of 3:2
(male:female). Previous studies found the mean age of disease
presentation to be in the fourth decade of life [92, 113, 117].
The mean age of disease presentation in our series was
40.2 years (range: 1 day–78 years old). Thus, the analysis of
our series confirmed the previously reported results. The cer-
vical spine followed by the thoracic spine was reported as the
most common localization of IMS [3, 85, 88, 89]. These find-
ings are also consistent with our analysis.

Previous studies addressing the clinical features and surgical
outcome of patients with IMS revealed sensory distur-
bance as the most common initial symptom [107]. Our results
show that patients with IMS suffer from sensory deficits as
often as from motor deficits, but we agree with Yang et al. on
the value of sphincter dysfunction as a late symptom [107].
Overall, patients with IMS seem to benefit from operation,
which is clearly shown by an improved postoperative neuro-
logical status in 86% of the patients. Previous studies on IMS
observed that patients with a longer symptom duration benefit
less from surgery due to chronic compression of the neuro-
mal tissue by the tumour [107]. In our review, we were not able
to confirm this hypothesis, since the analysis of the postoper-
ative outcome as a function of the duration of symptoms re-
vealed no significantly worse outcome for patients with a
symptom duration ≥10 years. In most of the cases, gross total
resection can be achieved easily [107]. In cases in which the
tumour is strongly adherent to the surrounding neuronal tis-
ue, subtotal resection should be considered in order to avoid
deterioration of the neurological status. In particularly compli-
cated cases, two-stage surgery provides a possible approach
towards better therapeutic results [91].

Conti et al. stated that IMS associates with NF; however,
several studies showed a prevalence of 0–2% in spinal tu-
mours [7, 70, 80, 103, 125]. Our review found NF in 11 of
166 cases (6.6%). These results reveal slightly higher rates of
NF in patients with IMS than previously described; however,
no firm association between NF and IMS was found.

IMS are frequently misdiagnosed as another tumour entity
because of the tumour location and its heterogenous appear-
ance in MRI diagnostics [113, 122]. Several series described
the MRI appearance of schwannomas as being iso/
hypointense in the T_1- and hyperintense in the T_2-weighted
images [1]. However, the T_1- and T_2-weighted appearance of
IMS varies among studies [107, 113]. The summary of these
studies in our review reveals that in most cases, IMS show a
similar MRI appearance as schwannomas. Specifically, in T_1-
weighted images, 35% of all cases appeared iso- or hypointense and in T_2-weighted images, 23.2% were
| Case No. | Reference | Localization | MRI |  |
|----------|-----------|--------------|-----|---|
| 1        | Penfield, 1932 [13] | C5 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 2        | Rasmussen et al., 1940 [14] | C4–7 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 3        | Roka, 1951 [15] | Cerv. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 4        | Rose, 1954 [16] | C5 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 5        | Riggs/Clary, 1957 [17] | C4/5 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 6        | Ramamurthi et al., 1958 [18] | T2 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 7        | Scott/Bentz, 1962 [19] | T3 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 8        | McCormick et al., 1964 [20] | L2 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 9        | Sloof, 1964 [9] | Cerv. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 10       | Mason/Keigher, 1968 [21] | T8–10 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 11       | Chigasaki/Pennybacker, 1968 [22] | T3 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 12       | Van Duinen, 1971 [23] | C3 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 13       | Fabres et al., 1972 [24] | T2/3 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 14       | Cambier et al., 1974 [25] | C2–4 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 15       | Wood et al., 1975 [26] | C3 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 16       | Schmitt, 1975 [27] | L1 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 17       | Isu et al., 1976 [28] | C1 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 18       | Kumar/Gulati, 1977 [29] | Cerv. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 19       | Vailati et al., 1979 [30] | T7–9 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 20       | Gegalian, 1979 [31] | T8/9 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 21       | Pardatscher et al., 1979 [8] | T2–8 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 22       | Shalit/Sandbank, 1981 [32] | C2-T2 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 23       | Guidetti, 1967 [33] | C3–5 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 24       | Lesoin et al., 1983 [35] | T12 – L1 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 25       | Lesoin et al., 1983 [35] | C3–7 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 26       | Rout et al., 1983 [36] | L1 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 27       | Kang/Song, 1983 [37] | C3–6 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 28       | Bouchez et al., 1984 [38] | C2–7 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 29       | Drapkin et al., 1985 [39] | C3–5 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 30       | Lesoin et al., 1986 [40] | T3–6 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 31       | Maruki et al., 1986 [41] | T7/8 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 32       | Ross et al., 1986 [4] | C2–T1 | Iso. | Hyper. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 33       | Adachi et al., 1990 [42] | C4/5 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 34       | Char/Cross, 1987 [42] | T3/4 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 35       | Garen et al., 1988 [43] | C3–6 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 36       | Hida et al., 1988 [44] | T8/9 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 37       | Okuda et al., 1988 [45] | Med.–C7 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 38       | Gorman et al., 1989 [46] | C5/6 | Mixed | Hyper. | n.a. | No | No | No | No |
| 39       | Sharma et al., 1989 [47] | C5 | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. | n.a. |
| 40       | Meisel et al., 1990 [48] | T9/10 | Hyper. | Hypo. | Homo. | No | Yes | Yes | No |
| 41       | Li/Holtas, 1991 [49] | C2 | Hypo./Iso. | Iso./Hypo. | Homo. | No | Yes | No | No |
| Case No. | Reference | Localization | MRI |
|----------|-----------|--------------|-----|
|          |           | Vertebra     | T1  | T2  | GA | CYS | OE | SYX |
| 43       | Herregodts et al., 1991 [50] | T2 | Hyper. | n.a. | Homo. | No | Yes | No |
| 44       | Jacquet et al., 1992 [51] | T12–L1 | n.a. | n.a. | Homo. | n.a. | n.a. | n.a. |
| 45       | Morimoto et al., 1992 [52] | T7–9 | n.a. | n.a. | Homo. | n.a. | n.a. | n.a. |
| 46       | Benini et al., 1993 [53] | T7–9 | n.a. | Hyper. | Minimal | No | No | No |
| 47       | Sekerci et al., 1993 [54] | C5/6 | n.a. | Iso. | Homo. | No | No | Yes |
| 48       | Radhakrishnan et al., 1993 [55] | T1–3 | n.a. | n.a. | Homo. | n.a. | n.a. | n.a. |
| 49       | C5/6 | n.a. | n.a. | Homo. | n.a. | No | Yes | No |
| 50       | C4–6 | n.a. | n.a. | Homo. | n.a. | n.a. | n.a. | n.a. |
| 51       | Nicoletti et al., 1994 [56] | C3–5 | Hyper. | Hypo. | Homo. | n.a. | No | No | No |
| 52       | Duong et al., 1995 [57] | T5–7 | Iso. | Iso. | Homo. | Yes | Yes | Yes |
| 53       | Melancia et al., 1996 [58] | T11–L2 | n.a. | n.a. | Homo. | n.a. | n.a. | n.a. |
| 54       | Lee et al., 1996 [2] | C5–T3 | n.a. | n.a. | Homo. | n.a. | n.a. | n.a. |
| 55       | C5 | n.a. | n.a. | Homo. | n.a. | n.a. | n.a. | n.a. |
| 56       | Botelho et al., 1996 [59] | C4–6 | n.a. | n.a. | Homo. | Yes | No | Yes |
| 57       | Innocenzi et al., 1996 [60] | C1–3 | Hypo. | Hyper. | Homo. | No | No | No |
| 58       | Bekar et al., 1997 [61] | C2–T1 | Hyper. | Hyper. | Homo. | Yes | No | No |
| 59       | Beşkonakli et al., 1997 [62] | T8 | Hyper. | n.a. | Homo. | No | Yes | No |
| 60       | Cotó et al., 1998 [63] | T4/5 | Hypo. | Iso. | Homo. | n.a. | n.a. | Yes |
| 61       | Kotil et al., 1998 [64] | T10/11 | n.a. | Hyper. | Homo. | n.a. | n.a. | n.a. |
| 62       | Hejazi/Hassler, 1998 [65] | T12–L1 | n.a. | n.a. | Homo. | n.a. | n.a. | n.a. |
| 63       | Binati et al., 1999 [66] | C6–T1 | n.a. | n.a. | Homo. | n.a. | n.a. | n.a. |
| 64       | Arellanes-Chávez et al., 2000 [67] | C2–5 | Iso. | Hyper. | Homo. | Yes | No | Yes |
| 65       | Riflaud et al., 2000 [3] | C1/2 | Hyper. | Hypo. | Homo. | No | No | Yes |
| 66       | Ogunjogo et al., 2000 [68] | C4–7 | n.a. | n.a. | Homo. | No | No | Yes |
| 67       | Kodama et al., 2000 [69] | C3–5 | Hyper. | Iso./Hypo. | Homo. | Yes | No | No |
| 68       | C1 | Hyper. | Hyper. | Circ. | Homo. | Yes | Yes | Yes |
| 69       | Patronas et al., 2001 [70] | T2 | Iso./Hyper. | Homo. | Yes | No | No |
| 70       | Kono et al., 2001 [71] | C2 | n.a. | n.a. | Homo. | No | No | No |
| 71       | Maira et al., 2001 [72] | C5/6 | Hypo. | Iso. | Homo. | n.a. | n.a. | n.a. |
| 72       | Sasaki et al., 2002 [73] | C3/4 | n.a. | n.a. | Homo. | No | No | Yes |
| 73       | Darwish et al., 2002 [74] | T3–8 | n.a. | n.a. | Heter. | No | No | Yes |
| 74       | Brown et al., 2002 [75] | O'Brien et al., 2003 [76] | T11–L1 | n.a. | Hyper. | Homo. | n.a. | n.a. | n.a. |
| 75       | Colosimo et al., 2003 [77] | C2 | Iso. | Hypo. | Homo. | n.a. | n.a. | n.a. |
| 76       | Panagiotopoulos et al., 2004 [78] | T6 | Hypo. | Homo. | No | No | No |
| 77       | Siddiqui/Shah, 2004 [79] | T9/10 | Hypo. | Homo. | No | No | No |
| 78       | Conti et al., 2004 [80] | Med.–C3 | Iso./Hypo. | Homo. | Heter. | No | No | Yes |
| 79       | C1 | n.a. | n.a. | Homo. | n.a. | n.a. | n.a. | n.a. |
| 80       | C4–6 | n.a. | n.a. | Homo. | n.a. | n.a. | n.a. | n.a. |
| 81       | Chavez-Lopez et al., 2004 [81] | C4–6 | Iso. | Homo. | No | No | Yes |
| 82       | El Malki et al., 2005 [82] | C1–6 | Hyper. | Homo. | Heter. | Yes | No | Yes |
| 83       | Amato et al., 2005 [83] | C4 | Hyper. | Homo. | No | No | Yes |
| 84       | Matsuyama et al., 2009 [84] | Kim et al., 2005 [85] | T8/9 | n.a. | Iso. | Homo. | No | Yes | No |
| Case No. | Reference                  | Localization | MRI |
|---------|----------------------------|--------------|-----|
| 89      | Kyoshima et al., 2005 [86] | T9/10        | Iso./Hypo. Iso. Circ. No No No |
| 90      | Shenoy/Raja, 2005 [87]    | C4–7         | Iso./Hypo. Hyper. Circ. No No Yes |
| 91      | Kahilogullari et al., 2005 [88] | T12–L2 | n.a. n.a. Heter. n.a. n.a. n.a. |
| 92      | Ho et al., 2006 [89]      | C5/6         | Iso. Hyper. Homo. No No No |
| 93      | Mukerji et al., 2007 [90] | C5–7         | Iso. Hyper. n.a. No Yes No |
| 94      | Hida et al., 2008 [91]    | C1/2         | Hypo. Iso. Heter. No Yes No |
| 95      | T2–6                      | n.a. n.a.    | Homo. No No No |
| 96      | Kim et al., 2009 [92]     | T5/6         | Hypo. Iso. Circ. No No Yes |
| 97      | Nicácio et al., 2009 [93] | C4–6         | Hyper. Hypo. Heter. No Yes Yes |
| 98      | Hayashi et al., 2009 [94] | T11–L1       | Hypo. Iso. Circ. Yes Yes No |
| 99      | Ohtonari et al., 2009 [95]| T12–L1       | Iso. n.a. Homo. Yes No No |
| 100     | Adam et al., 2010 [96]    | C2–5         | n.a. n.a. n.a. n.a. n.a. n.a. |
| 101     | T2–6                      | n.a. n.a.    | Homo. No No No |
| 102     | Lyle et al., 2010 [97]    | T2–Sac.      | n.a. Iso. Heter. No No No |
| 103     | Bernal-García et al., 2010 [5] | T1–5 | Iso. Hyper. Homo. No Yes No |
| 104     | C5–7                      | Hyper. Iso.  | Homo. No No No |
| 105     | Teo et al., 2011 [98]     | C5/6         | Hypo. Hyper. Homo. Yes Yes No |
| 106     | Ryu et al., 2011 [99]     | T6/7         | Iso. Hyper. Homo. No Yes Yes |
| 107     | Vij et al., 2011 [100]    | T10/11       | Hypo. Iso. n.a. No No No |
| 108     | Das et al., 2012 [101]    | C2/3         | Hypo. Hyper. n.a. No Yes No |
| 109     | Li et al., 2013 [102]     | T10/11       | Iso. Hypo. Heter. Yes No No |
| 110     | Lee et al., 1999 [103], Lee et al., 2013 [104]| C4–7 | n.a. n.a. Heter. n.a. n.a. n.a. |
| 111     | C5/6                      | n.a. n.a.    | Homo. n.a. n.a. n.a. n.a. |
| 112     | C5–7                      | n.a. n.a.    | Homo. n.a. n.a. n.a. |
| 113     | T1/2                      | n.a. n.a.    | Homo. n.a. n.a. n.a. |
| 114     | T6–8                      | n.a. n.a.    | Homo. n.a. n.a. n.a. |
| 115     | T7/8                      | n.a. n.a.    | Homo. n.a. n.a. n.a. |
| 116     | T7–10                     | n.a. n.a.    | Homo. n.a. n.a. n.a. |
| 117     | T8/9                      | n.a. n.a.    | Homo. n.a. n.a. n.a. |
| 118     | T9/10                     | n.a. n.a.    | Homo. n.a. n.a. n.a. |
| 119     | T10/11                    | n.a. n.a.    | Homo. n.a. n.a. n.a. |
| 120     | Eljebbouri et al., 2013 [105]| T7–9 | n.a. Hyper. Heter. Yes Yes No |
| 121     | Wu et al., 2011 [106], Yang et al., 2014 [107]| C6-T4 | Hypo./Iso. Hyper. Heter. Yes Yes No |
| 122     | C4–6                      | Hypo. Hyper. | Homo. No No Yes |
| 123     | C3–5                      | Iso. Iso.    | Homo. No No Yes |
| 124     | C6                        | Hypo. Hyper. | Homo. No No Yes |
| 125     | T3–5                      | Hypo./Iso.   | Hyper. Heter. Yes Yes No |
| 126     | C6/7                      | Hypo. Hyper./Iso. | Circ. Yes Yes No |
| 127     | T10–12                    | Hypo./Iso.   | Hyper./Iso. Heter. Yes No No |
| 138     | C3/4                      | Iso. Iso.    | Heter. No No No |
| 129     | C5/6                      | Hypo. Hyper. | Heter. Yes Yes No |
| 130     | T2/3                      | Iso. Iso.    | Homo. No No Yes |
| 131     | T9/10                     | Iso. Hyper.  | Homo. No No Yes |
| 132     | C1/2                      | Iso. Iso.    | Homo. No No Yes |
| 133     | C5/T1                     | Hypo. Hyper./Iso. | Heter. Yes Yes No |
| 134     | C4–6                      | Hypo./Iso.   | Hyper. Heter. Yes Yes No |
| Case No. | Reference              | Localization | MRI                  |
|---------|------------------------|--------------|----------------------|
| 135     | C5–7                   | Iso.         | Hyper./Iso.          | Heter. | No | No | Yes |
| 136     | T3                     | Iso.         | Iso.                 | Homo.  | No | Yes | No  |
| 137     | C3                     | Iso.         | Hyper.               | Heter. | No | No | Yes |
| 138     | T12                    | Iso.         | Hyper./Iso.          | Heter. | Yes| No | Yes |
| 139     | T6–8                   | Iso.         | Hyper./Iso.          | Heter. | Yes| No | Yes |
| 140     | T11                    | Iso.         | Iso.                 | Homo.  | No | No | No  |
| 141     | Yang et al., 2015 [108]| T11/12       | Iso.                 | Hypo.  | Heter. | Yes | No | Yes |
| 142     | Gupta et al., 2015 [109]| C3/4        | n.a.                 | Iso.   | Heter. | Yes | No | No  |
| 143     | Jagannatha et al., 2016 [110]| T11/12 | Hyper./Hypo.        | Hypo.  | Heter. | Yes | No | Yes |
| 144     | Sun et al., 2017 [111]| C1/2        | Iso.                 | Iso.   | Homo. | No | No | Yes |
| 145     | Nayak et al., 2017 [112]| T1–9       | Hyper.               | Hypo.  | Homo. | No | No | No  |
| 146     | Gao et al., 2017 [113]| T8/9        | Iso.                 | Hypo./Hyper.| Heter. | No | Yes | No  |
| 147     | T9/10                   | Hypo.        | Hypo.                | Heter. | No | Yes | No  |
| 148     | T10                    | Hypo.        | Hypo.                | Heter. | No | Yes | Yes |
| 149     | T4–6                   | Hypo.        | Hyper.               | Homo.  | No | No | No  |
| 150     | T10/11                  | Hypo.        | Hypo.                | Homo.  | No | No | No  |
| 151     | C6–T1                  | Hypo.        | Hypo./Hyper.         | Homo.  | No | Yes | Yes |
| 152     | C5/6                   | Hypo.        | Hypo./Hyper.         | Homo.  | No | Yes | No  |
| 153     | C4–7                   | Hypo.        | Hypo./Hyper.         | Homo.  | No | No | No  |
| 154     | Karatay et al., 2017 [114]| T12/L1       | Hypo.                | Hyper. | Homo. | No | No | Yes |
| 155     | Li et al., 2017 [115]| C3–5        | n.a.                 | n.a.   | n.a. | n.a. | n.a. | n.a. |
| 156     | Navarro Fernández et al., 2018 [116]| C6–7     | Iso.                 | Hyper. | Circ. | Yes | Yes | No  |
| 157     | Landi et al., 2018 [117]| T10/11     | n.a.                 | Hypo.  | Homo. | No | No | No  |
| 158     | Singh et al., 2018 [118]| T12–L2    | Hypo./Hyper.         | Hyper. | Heter. | Yes | No | No  |
| 159     | Wang et al., 2018 [119]| T8         | Hypo.                | Iso.   | Homo. | No | No | Yes |
| 160     | Shi et al., 2019 [120]| Cerv.       | n.a.                 | n.a.   | n.a. | n.a. | n.a. | n.a. |
| 161     | Dhake/Chatterjee, 2019 [121]| T10–12   | Iso./Hypo.           | Hyper. | Heter. | No | No | No  |
| 162     | T9/10                   | Hypo.        | Hyper.               | Circ.  | No | No | No  |
| 163     | Dai et al., 2019 [122]| C3/4       | Iso.                 | Hyper. | Minimal | No | Yes | No  |
| 164     | Sekar et al., 2019 [123]| C5–7       | Hypo.                | Hyper. | n.a. | Yes | No | Yes |
| 165     | Kelly et al., 2020 [124]| C4–T2     | Iso./Hypo.           | Hyper. | Heter. | No | No | Yes |

*MRI* magnetic resonance imaging  
*T1* T1-weighted images  
*T2* T2-weighted images  
*GA* gadolinium enhanced  
*CYS* cystic lesion  
*OE* oedema in T2-weighted images  
*SYX* tumour-associated syringomyelia  
*iso.* isointense  
*Hypo.* hypointense  
*Hyper.* hyperintense  
*Homo.* homogenous  
*Heter.* heterogenous  
*Circ.* circular  
*n.a.* information not available
1848

hyperintense. Interestingly, 1/5 of all cases associated with syringomyelia and in 20%, a perilesional edema was observed. The treated patient in our institution suffered from a perilesional edema, which showed a complete remission in the follow-up MRI after 4 months.

The pathogenesis of IMS is controversially debated among experts because of the absence of Schwann cells within the central nervous system (CNS) in healthy individuals [69]. Currently, there are six hypotheses regarding the origin of IMS: (a) conversion of pial mesodermal cells into neuroectodermal Schwann cells [126]; (b) migration and late neoplastic growth of ectopic Schwann cells during embryonal development [18, 30]; (c) origin from Schwann cells from the perivascular nerve plexus surrounding the blood vessels within the CNS [17, 27, 36, 127, 128]; (d) schwannosis in proximity to the anterior spinal artery [129]; (e) centripetal growth from a dorsal nerve root entry zone into the spinal cord [20, 21, 26, 128] and (f) result from imperfect regeneration of the spinal cord after mechanical trauma or chronic disease [130].

Although some association of proliferating vessels around the tumour [4, 32, 35, 68, 102], tumour connection to a nerve root [4, 27, 34, 43, 46, 52, 58, 68, 71, 76, 77, 84, 89, 99, 104, 107, 109, 115, 123] or chronic disease of the spinal cord could be observed in reported cases [39, 100, 107], it is still not possible to make a general statement regarding the pathogenesis of IMS. In our case, a tumour connection to the nerve root could be observed in the MRI of the cervical spine. This is why we rather support the hypothesis of centripetal growth from a nerve root entry zone into the spinal cord as a possible pathomechanism for development of IMS. However, this mechanism is not able to explain the formation of multiple IMS. The special subgroup of multiple IMS might have implications for the pathomechanism of IMS, but the available information do not allow a conclusions about differences in the pathogenesis of singular and multiple IMS.

As part of the preoperative examination and consultation of patients with intramedullary tumours, it is important to make a correct tentative diagnosis to ensure the best possible treatment. Since IMS are benign tumours of the spinal cord, their treatment might differ from other tumours, like spinal astrocytoma or ependymoma. Patients with IMS show a low rate of tumour recurrence. Even in cases with subtotal tumour resection, tumour recurrence is not necessarily observed [107]. In contrast, for patients with spinal ependymoma, the gross total resection is the gold standard to achieve the longest possible progression-free survival [131–134]. Therefore, complete removal of the tumour should be the goal of the surgery. Furthermore, it is unclear if patients with spinal astrocytoma benefit from gross total resection as patients with spinal ependymoma do [135–138]. Additionally, gross total resection is difficult to achieve in patients with spinal astrocytoma without causing a worse neurological outcome, which is why the primary goal of surgery is to spare the surrounding nervous tissue [139, 140]. Unfortunately, spinal astrocytoma and ependymoma are difficult to distinguish from IMS by use of MRI [107, 113, 141]. Therefore, it seems to be important to differentiate intramedullary tumours during surgery with the aid of intraoperative frozen sections in order to provide the patient with the best possible therapy [95, 104].

**Conclusion**

IMS are rare tumours of the spinal cord. One hundred sixty-six cases have been reported so far, including the here reported case. IMS are more frequently found in male patients; the mean age of disease presentation is the fourth decade of life. The most common localization of IMS is the cervical spine, followed by the thoracic spine. Although several explanations regarding the pathogenesis of IMS have been proposed, it is still not possible to make a general statement regarding the pathogenesis of these tumours, especially for the subgroup of patients with multiple IMS. In our study, no firm association between NF and IMS was found.

Patients suffering from IMS present in most of the cases with sensory and motor deficits; sphincter dysfunction seems to be a late symptom. Due to heterogenous imaging patterns in MRI, it is difficult to preoperatively differentiate an IMS from other intramedullary tumours. Therefore, intraoperative frozen section might be useful to determine the tumour entity and the best suited surgical strategy. Overall, patients with IMS seem to benefit from operation; in most of the cases, gross total resection can be achieved easily. Nevertheless, further multicentre studies are necessary to elucidate the pathomechanism leading to IMS formation and to determine strategies for the best clinical care for these patients.

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**Code availability**  Not applicable.

**Author’s contributions**  All mentioned authors contributed to the study conception and design. Literature search and data collection were performed by VMS. Data analysis and writing of the first manuscript draft were performed by VMS and BN. All authors commented on previous versions of the manuscript and approved the final manuscript.

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**Data availability**  The authors declare that the data supporting the findings of this study are available within the article.

**Compliance with ethical standards**  

**Conflict of interest**  The authors declare that they have no conflict of interest.
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