Case Report

Intramedullary holocord mature teratoma in an adult- case report and review of the literature

Nikolaos Saridakis\textsuperscript{a}, Christina Koumantzia\textsuperscript{b}, Sylwia Libard\textsuperscript{c,d}, Niklas Marklund\textsuperscript{e}, Andreas Eleftheriou\textsuperscript{a,*}

\textsuperscript{a} Department of Neurology in Linköping and Department of Biomedical and Clinical Sciences, Linköping University, Linköping, Sweden
\textsuperscript{b} Primary Health Care Center in Tannefors and Department of Medical and Health Sciences, Linköping University, Linköping, Sweden
\textsuperscript{c} Department of Immunology, Genetics and Pathology, Uppsala University, Sweden
\textsuperscript{d} Department of Pathology, Uppsala University Hospital, Sweden
\textsuperscript{e} Lund University, Skane University Hospital, Department of Clinical Sciences Lund, Neurosurgery, Lund, Sweden

A R T I C L E   I N F O

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1. Introduction

Teratomas are tumours of germ cell or dysembryogenic origin that result from ectopic growth of two or more totipotent cell lines (ectoderm, mesoderm, endoderm), while true teratomas are defined as tumours derived from all three germ layers (tridermal) [1,2]. Nonomura et al. in 2002 published a case of a bidermal tumour, defined as mature teratoma and noted that such cases of bidermal teratomas had even been reported before. The presence of only two germinal components does not necessarily rule out the diagnosis of teratomas because cell-components of one or two germ layers tend to overgrow the others, so that the total number of germ layers may be difficult to ascertain [3]. Histologically, teratomas can be divided into three categories: mature, immature, and malignant [4]. Dermoid cysts have previously been considered a kind of mature cystic teratoma histopathologically including elements from 2 germ layers (ectoderm and mesoderm) [5]. Others define dermoid cysts as a separate entity originated entirely from the ectoderm and thus if a dermoid cyst contains adipose tissue which derives from the mesoderm, it would be termed as teratoma [6]. Dermoid cysts are usually composed of keratinized squamous epithelial lining covered by connective tissue, islands of dermis containing hair follicles and sebaceous glands [5].

Both teratomas and dermoid cysts occur rarely in the spinal cord with teratomas accounting for only 0.2 % - 0.5 % and dermoid cysts for 0.8–1.1 % of all spinal cord tumours [3,7–9]. Intraspinal teratomas are rarely found in adults while most dermoid cysts, although mostly congenital, have been diagnosed during 2nd and 3rd decades of life probably because of slow growth and absence of symptoms [3,5]. These lesions are predominantly located in the lumbo-sacral region and they can be divided into intramedullary, intradural extramedullary or extradural [1,10]. Regarding tumours referred in the literature as intramedullary teratomas, there was non-statistically significant sex predominance according to the last large systematic review [8] while other reviews suggest that they may affect women more often among adults and boys among children [8,11]. Regarding lesions referred as dermoid cysts, a slight male predominance has been observed [5]. Whether mature teratomas include dermoid cysts is though still controversial. Epidemiological characteristics of such tumours in adults are presented with some discrepancy due to the diverse nomenclature and definition, but there are no differences in therapeutic approach and surgical treatment [10].

Herein, we present a unique case of a holocord-panmedullary expansive mature teratoma expanding from the medulla oblongata to L2 in an adult male. We also reviewed the literature regarding
Table 1
All the intramedullary dermoid cysts. The cases with * refer to ruptured dermoid cysts.

| Author, Year [Ref No.] | No. Cases | Sex | Age | Extension | Surgical removal | Outcome |
|-------------------------|-----------|-----|-----|-----------|-----------------|---------|
| Harriehausen H, 1909 [20] | 1 | F | 23 | L1 | Not found | |
| Mixter, 1932 [21] | 1 | M | 23 | C2-C5 | Not found | |
| Love and Kernohan, 1936 [22] | 1 | M | 36 | Lumbosacral | Not found | |
| List, 1941 [23] | 1 | M | 19 | T1-T5 | Not found | |
| Kooster and Rapids, 1942 [24] | 1 | M | 22 | C7-T4 | Not found | |
| Woods and Pimenta, 1944 [25] | 1 | F | 46 | T12-L3 | Not found | |
| Thurel et al, 1952 [26] | 1 | M | 40 | T5 | Not found | |
| Tyrus and Pennybaker, 1956 [27] | 1 | M | 26 | T12 | Not found | |
| Roth et al, 1966 [28] | 1 | M | 62 | T8-T12 | Partial 6 years after the first admission to hospital because of clinical deterioration. (The patient had refused surgery initially) | Deterioration with secondary to immobility complications and death 5 months after surgery |
| Gagliardi and Ferrari, 1969 [29] | 1 | M | 25 | T12-L2 | Partial | Probably complete recovery |
| Arseni et al, 1977 [30] | 1 | F | 24 | T12-L2 | Partial | Improvement |
| 2 | F | 19 | T11-T12 | Radical | Major improvement |
| 3 | F | 24 | C5-C6 | Partial (+Rx therapy) | Major improvement (Rx therapy had influence on pain relief and not on the tumour) |
| Graham et al, 1988 [31] | 1 | M | 34 | Conus (T12-L2) | Partial | No improvement |
| Lunardi et al, 1989 [32] | 1 | F | 48 | T8-T9 | Partial | Improvement |
| 2 | M | 43 | T12-L2 | Partial | Major improvement (reoperation 12 years later) |
| 3 | F | 21 | T2-T5 | Partial | Improvement |
| 4 | M | 25 | T9-L3 | Radical | Improvement |
| Bani et al, 1992 [33] | 1* | M | 27 | T11-L1 | Probable radical | Improvement |
| Cavazzani et al, 1995 [34] | 1* | M | 63 | Conus | Partial with recurrence and rupture causing hydrocephalus 19 years later. | After reoperation with radical removal + VP shunt Major improvement. |
| 2* | M | 55 | T12-L1 | Radical 12 years after shunt operation because of hydrocephalus which was probably the result of the undetected cyst. | Major improvement |
| Calabro, 2000 [18] | 1* | M | 33 | L1 | Probably radical | Major improvement |
| 2* | M | 44 | Lumbar enlargement | Partial | NM |
| Falavigna et al, 2001 [35] | 1 | F | 48 | T11-T12 | Partial | Major improvement |
| Karadag et al, 2002 [36] | 1* | M | 55 | T12-L1 | NM | NM |
| Garg A. et al, 2003 [33] | 1* | M | 32 | Conus | Not specified | Major improvement |
| 2* | M | 34 | Conus | Probably partial | NM |
| Goyal et al, 2004 [37] | 1* | M | 32 | Conus | Not found | Not found |
| Najjar et al, 2005 [38] | 1 | F | 31 | T7-T8 | Probably partial | No change |
| Jidal et al, 2005 [39] | 1* | NM | 40 | Conus | Partial | Improvement |
| Cha et al, 2006 [40] | 1* | M | 44 | L3-L5 | Not specified | Improvement |
| Gopen et al, 2006 [41] | 1* | M | 35 | Conus | Not specified | Complete recovery |
| Kasliwal et al, 2007 [42] | 1* | M | 26 | Conus | Probably radical | Total recovery |
| Muthukumar et al, 2007 [43] | 1 | M | 19 | Conus | Radical | Improvement |
| Ogden et al, 2007 [44] | 1 | M | 23 | C7-T1 | Partial | Improvement |
| Vyas et al, 2010 [19] | 1* | M | 20 | Conus | Probably partial at the age of 20 with recurrence and rupture diagnosed at the age of 35. NM if reoperation. | Recurrence with complication. |
| Patankar and Sheth, 2012 [45] | 1 | F | 18 | C3-T2 | Partial | Improvement |
| Signorelli et al, 2013 [46] | 1 | M | 65 | C2-C5 | Partial | Deterioration |
| Garg K., 2014 [5] | 1* | M | 19 | L2-L3 | Radical | No change |
| 2* | M | 23 | L1-L2 | Partial | Improvement |
| 3* | M | 31 | Conus | Partial | No change |
| Shuxheng and Yazhou et al, 2014 [10] | 1 | F | 24 | T12-L3 | Partial | Improvement |
| 2 | M | 20 | T10 | Radical | Complete recovery |
| Seerangan et al, 2018 [47] | 1 | F | 18 | C5-C7 | Radical | Improvement |
| 1 | F | 18 | Conus | Partial | No deterioration |
intradural mature teratomas in adults (Table 1 and 2) in order to approach the epidemiology and therapeutic evaluation. There are previous reviews regarding intraspinal teratomas, the most recent published in 2009, including all the cases of mature and immature intradural teratomas. Because of the above mentioned discrepancy regarding definition and nomenclature of spinal teratomas and dermoid cysts, we undertook a systematic review of all the intradural mature teratomas and intradural dermoid cysts in adults. We independently searched PubMed (until December 2019) using the following free text terms: “intradural mature teratoma”, “intradural dermoid cyst”, “intraspinal teratoma”, “intraspinal dermoid cyst”, “spinal teratoma”, “spinal dermoid cyst”, “teratoma of the spinal cord/spine”, “dermoid cyst of the spinal cord/spine”, “holorocord”, “panmedullary”. We included case reports, abstracts and articles in English, one in Italian and one in French. All results from the electronic searches were compiled in a reference manager program (Endnote X7) and all duplicated citations were eliminated. The following data were collected: (1) publication details such as title, authors, and other citation details, (2) patient data such as age and sex (3) details of tumours’ location, (4) data of surgical approach, (5) follow-up data regarding recovery and outcome. All in all, we reviewed more than 90 publications, which are cited in this article and finally found 45

### Table 2

All the intradural mature teratomas. Abbreviations: M = male, F = female, NM = not mentioned. Comments: Age in years is referred to patient’s age when the diagnosis was set.

| Author, Year [Ref No.] | No. Cases | Sex | Age | Extension | Surgical removal | Outcome |
|-------------------------|-----------|-----|-----|-----------|------------------|---------|
| Hosoi et al, 1931 [49]  | 1         | M   | 24  | Conus     | Partial          | Complete recovery |
| Dereymacker et al, 1954 [50] | 1   | F   | 43  | C5-T2     | Radical          | Improvement       |
|                        | 2         | M   | 34  | Conus     | Partial          | Minor improvement |
| Slooff et al, 1964 [51]  | 1         | M   | 20  | T11-L1    | Radical          | Improvement       |
|                        | 2         | M   | 67  | T11       | Autopsy          | –                   |
| Rewcastle et al, 1965 [52] | 1   | F   | 34  | T10       | Partial          | Minor improvement |
| Ikabay et al, 1965 [53]  | 1         | F   | 65  | Conus     | Partial          | Not found         |
| Hannebout et al, 1965 [54] | 1   | M   | 47  | Conus     | Radical          | Major improvement |
| Enstrom et al, 1977 [55] | 1         | M   | 36  | T11-L1    | Partial          | Improvement       |
| Rosenbaum et al, 1978 [56] | 1   | M   | 49  | T9        | Radical          | Improvement       |
| Besel et al, 1979 [57]  | 1         | F   | 22  | T11       | Radical          | Not found         |
| Garrison et al, 1980 [58] | 1   | M   | 23  | Conus     | Radical          | Complete recovery |
| Padovani et al, 1982 [59] | 1   | F   | 21  | C6-T1     | Partial          | Improvement       |
| Padovani et al, 1983 [60] | 1   | F   | 33  | L1-L3     | Radical          | Minor improvement |
| Conti et al, 1984 [61]  | 1         | F   | 24  | Conus     | Partial          | Not found         |
| Giacomini et al, 1986 [62] | 1   | M   | 31  | Conus     | Radical          | Improvement       |
| Pelissou-Guyotat et al, 1988 [63] | 1 | M | 33 | Conus | Partial | Improvement |
| Nocletti et al, 1994 [64] | 1         | M   | 47  | T12-L4    | Partial          | Improvement       |
| Caruso et al, 1996 [65]  | 1         | M   | 41  | Conus     | Radical          | Improvement       |
| Al-Sarraj et al, 1998 [66] | 1   | M   | 35  | Conus     | Partial          | Improvement       |
| Poeze et al, 1999 [15]  | 1         | M   | 23  | T12-L1    | Partial          | Improvement       |
| Arai et al, 2000 [67]   | 1         | F   | 43  | Conus     | Partial          | Improvement       |
| Ak et al, 2006 [68]    | 1         | F   | 43  | C2-C3     | Partial          | Improvement       |
| Kabilogiullari et al, 2006 [69] | 1 | F | 42 | Conus | Radical | Improvement |
| Caruso et al, 2006 [70]  | 1         | F   | 40  | Conus     | Partial          | Improvement       |
| Makary et al, 2007 [71]  | 1         | F   | 46  | C1-C2     | Radical          | Improvement       |
| Mut et al, 2007 [72]   | 1         | F   | 34  | L1-L2     | Radical          | Improvement       |
| Mohindra et al, 2008 [73] | 1   | M   | 35  | Conus     | Radical          | Major improvement |
| Benes et al, 2009 [74]  | 1         | F   | 52  | Conus     | Partial          | Improvement       |
| Arvin et al, 2009 [75]  | 1         | M   | 34  | C4-T6     | Partial          | Complete recovery |
| Ghostine et al, 2009 [76] | 1   | F   | 65  | C1        | Partial          | Minor improvement |
| Ijiri et al, 2009 [77]  | 1         | F   | 68  | Conus     | Radical          | No improvement    |
| Sharma et al, 1989—2009 [78] | 1 | F | 51 | T10-L2 | Not specified. Radical in 15/20 spinal teratomas | Not specified. |
| Jian et al, 2010 [79]  | 1         | M   | 18  | Conus     | Radical          | Complete recovery |
| Li et al, 2013 [80]    | 1         | F   | 22  | T12-L2    | Radical          | Improvement       |
| Turan et al, 2016 [81]  | 1         | M   | 48  | Conus     | Partial          | Improvement       |
| Asan et al, 2016 [82]  | 1         | F   | 29  | Thoracolumbar junction | Partial | Major improvement |
| Wang et al, 2016 [83]  | 1         | M   | 21  | L4-L5     | Partial          | NM                |
| Mohammadi et al, 2017 [84] | 1 | M | 18 | L2-L3 | Partial | Improvement |
| Guadarrama-Ortiz et al, 2017 [85] | 1 | F | 59 | C6-T1 | Radical | Improvement |
| Diyora et al, 2018 [86] | 1         | M   | 40  | C4-C7     | Partial          | Improvement       |
| de Oliveira et al, 2019 [87] | 1 | F | 35 | L2-L4 | Partial | Improvement |
| Saridakis et al, 2020 | 1         | M   | 36  | Holocord/Panmedullary | Partial | Improvement but with remaining neuropathic pan |
cases of intramedullary dermoid cysts and 56 intramedullary mature teratomas in adults from 1909, that is to say 102 cases including ours. In articles in which both the title and the whole text did not contain the term “intramedullary”, (instead e.g. spinal, intraspinal or intradural) we evaluated the history, MRI images and location before including these lesions in our Table.

After having created two tables, one for mature teratomas and one for dermoid cysts, we carried out a univariate statistical analysis regarding the following parameters: age of initial detection, gender, location, both separately for each table and totally for all the cases. Cases with non-mentioned data were excluded from the analysis.

To our knowledge, this is the second presentation of a histopathologically verified mature teratoma extending throughout the entire rostrocaudal extent of the spinal cord in an adult after that one presented by Nonomura et al. in 2002 and the first systematic review of the literature including all the intramedullary teratomas and dermoid cysts from 1909 [3]. Another one panmedullary lesion in an adult was reported by Sharma et al. in 2009 and it was suggested to be a dermoid cyst according to MRI, but that was not histopathologically verified as the patient refused the surgery [12].

2. Case report

A Middle Eastern Caucasian man presented with a family history free of neurological disorders and congenital anomalies in the spinal cord as well as a symptom-free childhood. Paresthesias with radiation to the right leg and foot were the first symptoms in early adulthood. Spasms and pain, of short duration and low intensity, in the lower extremities, trunk and lower spine area were other manifestations. No medical evaluation was performed until progression of symptoms with involvement of the upper chest and abdomen and the development of urination difficulties without incontinence, at the age of 36.

Brain Magnetic Resonance Imaging (MRI) demonstrated a lipoma-like tumour in the dorsal medulla oblongata (Fig. 1). Cervical spine Computed Tomography (CT) revealed no pathological findings. A whole spine MRI showed intramedullary dilated tortuous vessel formation in the cervical and thoracic region with associated hyper-intensity in T1 and T2 weighted images. Contrast-enhanced sequences detected no paraspinal soft tissue masses. The initial radiological interpretation was that these findings were consistent with intramedullary cavernoma, vascular malformation or haemorrhage of the intramedullary spinal cord cavernous angioma. Antidepressant medication and analgesics were introduced for treatment of neuropathic pain with good initial efficacy, but later supplementary treatment with carbamazepine and pregabalin became necessary.

At the age of 41, self-catheterisation was needed due to urinary retention, and baclofen medication due to lower limb spasticity was started. The clinical deterioration regarding increased spasticity in lower extremities, gait difficulties and balance problems prompted a new brain and spine MRI that revealed expansion of the spinal canal caused by a lipoma–like lesion that was contrast-enhanced at the periphery. At the L1-L2 level, the process filled the entire spinal canal. There was also a lesion that measured 4–5 mm dorsally in the medulla oblongata, at the level of the foramen magnum, with the same signal pattern as the one at the L1-L2 level. Moreover, the brain parenchyma and the ventricular system had normal appearance and signal intensity. After an additional six months, the lesions had not progressed on investigation with a follow-up MRI (Fig. 1). The risks of an operation outweighed the possible benefits while symptom-treatment focused against pain and spasticity had a good effect at that time. Preoperatively, the electroneurogram and electromyogram revealed signs of a sensorimotor affect including posterior column affection at the thoracic and lumbar level. This was in line with the neurological status, which showed affected tactile and vibratory sensation in both lower extremities and spastic tetraparesis that was more pronounced in the lower extremities (3 of 5 in muscle strength) than in the upper extremities (4 of 5 in muscle strength) and with right dominance. The daily medication before the surgery included baclofen, morphine, clorozaxon, paracetamol, gabapentin, pregabalin and duloxetine.

At the age of 43, due to acute worsening of patient’s neurological status including cauda equine syndrome and pain refractory to treatment, the patient was operated at Uppsala University Hospital, using laminectomy and intraoperative neurophysiological monitoring. Following a Th9-L2 laminectomy, a midline durotomy was performed and a swollen cord throughout the extent of the dural opening was observed. At the upper part, the external surface of the cord had a normal appearance while in the lower part it had the appearance of a yellowish lipoma formation. A midline incision in the cord was made at the transition of the normally appearing cord and the part with altered appearance. Tissue of variable configuration was removed, including frequent calcifications and tufts of hair. Cauda equine nerve roots could be observed within the lesion caudally and small remnants at the conus were not removed. During the resection, sensory evoked potentials could not be interpreted. The motor evoked potentials (MEPs) were intact, but became gradually decreased on the left side. At the end of the resection, the MEPs then returned to baseline. Since the indication for surgery was deterioration of the cauda equine function, we did not extend our resection cranially. The dura was closed with continuous sutures, and the muscle layers and subcutaneous tissue were closed by interrupted sutures. The skin was then closed by continuous sutures. No drain was used. Following bedrest for 48 h, the patient could be mobilized and the post-operative course was uneventful.

Postoperatively, the pain in the lower extremities decreased by 75%. Severe hyperesthesia and allodynia in the right arm and trunk (T4-T10) were unaltered. The gait was insecure and wide-based, but the patient could walk better than preoperatively. The proximal and distal muscle strength of the upper and lower limbs was good and symmetric.

Post-operative MRI showed complete resection of the tumour in the operated area (Fig. 2). The histopathologic diagnosis corresponded with a mature teratoma as all three germ layers were represented within the lesion including fibro-adipose tissue, brain tissue, nerve fascicles, skin with skin appendages and adenoid prostate tissue. All components were mature with no signs of atypia or anaplasia. The histopathologic features of the lesion are visualised in Fig. 3.

The patient was not considered a candidate for oncological treatment since mature teratoma is insensitive to both chemotherapy and radiotherapy. In order to exclude testicular cancer as the origin of the tumour, ultrasonography of the scrotum was performed and it was normal. Furthermore, S-PLAP (placental alkaline phosphatase), a tumour marker for seminoma, was within normal limits. Supplementary investigation with CT of the thorax and the abdomen showed no suspected signs of metastases. Because of an elevated Prostate-Specific Antigen (PSA) value (at 5.6 μg/L), a prostate biopsy was performed and it was normal. Urodynamic testing indicated a possible lower motor neuron lesion. The diagnosis of neurogenic micturnition disorder was made. Continued clean, intermittent self-catheterisation was recommended. PSA elevation was probably connected with the teratoma itself, which was assessed to be PSA-positive.

The post-operative neurological rehabilitation and follow-up was taken over by the Neurology Department of Linköping University Hospital, Sweden because the patient moved to Linköping. About eight months after the operation and despite the extended per oral medication as well as physiotherapy with Transcutaneous Electrical Nerve Stimulation, the patient experienced a deterioration of hyperesthesia and neuropathic pain in the lower extremities. Furthermore, despite per oral medication refractory pain and allodynia was noticed, and a new MRI of the whole spine was performed without showing any progress, which was in line with the stable neurological status (Fig. 2). That did not prompt any further surgical measurements and thus the patient was assessed as a candidate for advanced therapy with implantation of an intrathecal pump (SynchroMed II pump) for administration of bupivacaine and morphine, which resulted in a pain reduction of about 80%.

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Two years later, new deterioration of the clinical image due to the dysfunction of the pump which had probably been occluded, led to reoperation and a change of the whole pump system with good clinical effect.

3. Discussion

The origin of spinal cord teratoma is controversial. Three theories have been proposed: a) spinal teratomas constitute an abortive “twin” formation, b) cystic teratomas represent the ependymal diverticulum of the central canal of the spinal cord which, however, does not explain the presence of tissue originated from mesoderm, c) the most commonly accepted theory is that teratoma originates from multipotential germinal cells that are misplaced in early embryonic development [3].

Spinal dermoid cysts are mostly congenital and rarely acquired. The congenital ones rise from inclusion of ectodermal elements into the spinal canal at the time of neural tube closure between 3 and 5 weeks of embryogenesis, while acquired dermoid cysts can occur after the introduction of dermal elements into the spinal subarachnoid space due to lumbar puncture, surgery or trauma. [13].

The analysis of our database, Table 1 and Table 2, is presented in Table 3 including the results obtained from the combined data. Our results are in line with previous literature apart from the male: female ratio among dermoid cysts which is higher according to our results.

Teratoma and dermoid cyst should be taken into consideration in differential diagnosis of intramedullary lesions. Neuroimaging is helpful, but definitive diagnosis is made by histopathological examination [14,15]. Neuroradiological findings in CT and MRI may include mixed intensities revealing different tissue characteristics, calcification of the lesion and associated vertebral anomalies. As far as we know, it is also recommended to exclude germ cell tumours as the origin of such middle line neoplasias, as well as to exclude metastases in the thorax and the abdomen. Treatment of intramedullary teratoma is surgical, and radical removal of the tumour should be the aim whenever possible, aided by intraoperative electrophysiological monitoring [16]. Subtotal resection is a valid alternative to radical tumour removal when neurological function is at risk. Recurrences were most commonly encountered in immature or malignant teratomas [15,17]. Additionally, an intraoperative complication during surgical removal of dermoid cysts includes the spillage of cystic components which may cause aseptic chemical meningitis. Thus, in the case of a large cystic component, needle aspiration of the cystic contents prior to debulking and/or resection should be taken into consideration.

The overall prognosis of adult patients with intramedullary mature teratoma or dermoid cyst is relatively good following surgical resection, with most patients showing either stabilization or improvement [8,9]. Long-term follow-up should include repeated office visits and control MRI imaging to assess for any possible recurrence or tumour progression, especially in cases of residual tumour [15]. Given that both mature teratomas and dermoid cysts grow relatively slowly and are benign, there is no role for adjuvant radiotherapy or chemotherapy [9].

Herein, we presented a rare case of a holocord intramedullary mature tridermal/true teratoma in an adult, which was the second reported case of a histologically verified holocord intramedullary mature teratoma in adults. Surgical resection was performed at the T9-L2 level.
Fig. 2. Post-operative MRI of the whole spine showing the holocord expansive mature teratoma expanding from the medulla oblongata to L2.

Fig. 3. Photos of a sample from this intramedullary mature teratoma. In A, hematoxylin - eosin (HE) stained section representing fibro-adipose tissue with a cyst lined with keratinizing squamous epithelium, surrounded by sebaceous glands. In B and C island of brain parenchyma within fibro-adipose tissue stained with HE in B and visualised immunohistochemically (IHC) applying antibody (Ab) towards glial fibrillary acidic protein (polyclonal, DAKO-Agilent) in C. In D and E adenoid prostate tissue within fibro-adipose tissue stained with HE in D and IHC stained applying Ab towards prostate-specific antigen (polyclonal, Dako-Agilent) in E. Bars A 500 μm, B-E 100 μm.
while the rest of the tumour remained untouched due to the fact that the symptoms included mostly lower extremities and because of the acute worsening, with cauda-equine syndrome. The clinical image improved significantly postoperatively, even including pain reduction up to 75%. Advanced therapy with an intrathecal pump was selected to treat refractory, despite per oral medication, neuropathic pain while the neurological status was stable. An alternative diagnosis and pathophysiological mechanism in our case could be a cystic intramedullary mature teratoma in conus medullaris which ruptured locally, and fatty droplets disseminated and extended rostrally up to the medulla oblongata. This alternative is however not supported by our MRI images and histopathological examination. Nevertheless, the minor possibility of a cyst content gradually migrating upwards into cervical region from a localized lumbar lesion must be considered. This possibility could only be ruled out by a tissue biopsy or radical surgery, for which there was no clinical indication at that moment. Moreover, the patient had neurogenic pain in the upper extremities strongly arguing against an effect of central canal cyst content that with a high probability should remain asymptomatic.

Searching PubMed for ruptured intramedullary teratoma or dermoid cyst, only 17 cases of ruptured dermoid cysts were found and no case of ruptured teratoma. However, all the authors describe ruptures with dissemination of fatty droplets to other parts of spinal cord more rostrally or into the ventricles. Additionally, all the ruptured “dermoid cysts” included adipose tissue which derived from mesoderm and thus according to some authors, as mentioned in the introduction, should be considered as mature teratomas (cystic). It is also significant and interesting that all the cases of ruptured intramedullary “dermoid cysts” referred to males. We found only one case of a ruptured “dermoid cyst” in a female which was extramedullary and was not included in our Table 2 [18].

Although dermoid cysts and mature teratomas are benign, accumulation of secretions can cause size growth which can lead to rupture either spontaneously or following surgery or trauma. A rupture can occur either into the subarachnoid space leading to obstructive hydrocephalus or chemical meningitis, or more rarely into the central canal, typically remaining asymptomatic. Syringohydromyelia is a rare concomitant finding in cases of spinal dermoid cysts, observed mostly in cases of ruptures into the central canal. Thus, rupture should be suspected in cases of spinal dermoid cyst along with syringohydromyelia and, if indicated, a screening study of the spine and brain is recommended with MRI. This is a rare event and only a few cases have been reported in literature [5,13,19].

4. Conclusion

Teratomas are a rare diagnosis in clinical practice. Neuroimaging is helpful, but definitive diagnosis is made by histopathological examination. Radical or subtotal resection should be attempted, using neurophysiological monitoring, with the aim of achieving a cure or at least symptom improvement. The rarity of our case presentation, a holocord intramedullary mature teratoma extending from the medulla oblongata to L2 in an adult, motivated an extended review of the literature. We noticed the discrepancy in definition and nomenclature of mature teratomas and dermoid cysts, which makes the epidemiological study of such lesions difficult. As part of differential diagnostic process for our case, we reviewed all publications of ruptured intramedullary dermoid cysts and found out that all cases were observed in males. The pathophysiology explaining this novel finding of male over-representation is unknown. This observation should be addressed in future studies.

5. Availability of data and material

We used the data from our Cambio COSMIC Healthcare System, which is a digital comprehensive healthcare system installed in all clinics in our region. Radiological material was obtained through Sectra Image Display System 7.

Authors’ contributions

Andreas Eleftheriou was the neurologist who performed the clinical and neurological evaluation. Andreas Eleftheriou, Christina Koumantzia and Nikolaos Saridakis were the major contributors in writing the manuscript. Niklas Marklund and Konstantin Salci were the neurosurgeons who performed the operation. Sylvia Libard performed the histopathological analysis.

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Declaration of Competing Interest

The authors declare that they have no competing interests.

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Table 3

Analysis of our database from Table 1 and Table 2.

|                      | Intramedullary Dermoid cysts (table 1) | Intramedullary mature teratomas (table 2) | Combined data |
|----------------------|----------------------------------------|------------------------------------------|---------------|
| Median age in years  | 26 (18–65)                             | 34 (18–68)                               | 27 (18–68)    |
| (range)              |                                        |                                          |               |
| Percentage males     | 70 %–30 % (more than 2:1)              | 54 %–46 % (about 1:1)                    | 61 %–49 % (3:2.5) |
| Percentage females   |                                        |                                          |               |
| Most common location | Lumbosacral region (60 %)               | Lumbosacral region (68 %)                | Lumbosacral region (65 %) |
|                      |                                        |                                          |               |
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