Acute paraplegia due to thoracolumbar schwannoma following trauma: A case report and literature review

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Abstract. Spinal schwannomas account for one third of primary spinal neoplasms. Clinical presentation is related to the tumor location. An atypical case of acute paraplegia following a fall, on the ground of a thoracolumbar schwannoma, without intratumoral hemorrhage, in a previously asymptomatic patient is reported. A 58-year-old male patient presented with acute paraplegia, and urinary and bowel incontinence, following a fall. The patient had no previous history of back and/or leg pain or neurological symptoms. Magnetic resonance imaging revealed a subdural mass, as well as a fracture of the right T12-L1 facet joint and the right transverse process. The patient underwent emergency T11-L1 wide laminectomy, exploration of the subdural space and T10-L2 posterolateral transpedicular stabilization and fusion. An intradural, extramedullary mass, causing severe cord compression, was found and excised. Pathology revealed schwannoma, without intratumoral hemorrhage. The patient recovered completely 6 months postoperatively. To the best of our knowledge, this is the first report of spinal intradural schwannoma causing sudden paraplegia in a previously asymptomatic patient in the setting of trauma, without intratumoral hemorrhage. Emergency canal decompression and complete excision of the tumor represent the optimal management of such cases.

Introduction

Nervous sheath tumors account for approximately 25% of tumors of the intradural-extradural space, while approximately 65% of them are schwannomas (1). Schwannomas represent slow-growing benign tumors arising from Schwann cells of the nerve sheaths of peripheral nerves and believed to originate from embryonic neural crest cells (1,2).

The presenting symptoms depend on the tumor's location and the degree of the spinal cord or the nerve root compression. Patients usually complain about pain, motor deficits, paresthesia and numbness. Symptoms are usually slowly progressive, with some cases being asymptomatic and accidentally diagnosed by imaging. Acute paraplegia is an extremely rare presenting symptom usually associated with intratumoral hemorrhage with or without related trauma (2). Furthermore, the growth pattern of spinal schwannomas is not well known. Therefore, although radical excision is recommended for symptomatic tumors, the optimal treatment for the asymptomatic ones remains unclear (2).

A case of 58-year-old male, suffering from thoracolumbar schwannoma presenting with acute paraplegia, after a fall is described. Histopathology did not reveal hemorrhage.

Taking into account the existing literature, the present represents the first case of acute paraplegia, following trauma, due to a thoracolumbar schwannoma, without intratumoral hemorrhage, in a previously asymptomatic patient.

Case report

A 58-year-old male, with unremarkable medical history, was admitted to the University Hospital of Heraklion (Heraklion, Greece) due to acute paraplegia following a fall from a tree (a height of 3 m).

The patient was oriented, afebrile (36.5°C) and hemodynamically stable (blood pressure=130/95 mmHg, heart rate=85 beats/min). He had no prior history of back pain or any other symptoms.

On admission, motor examination revealed grade 0/5 power in both lower limbs. Sensation of lower limbs was
impaired in both: light touch, as well as pin prick, while patellar and Achilles reflexes were absent. Furthermore, there was urinary and bowel incontinence.

Emergency computer tomography (CT) scan showed fracture of the right T12-L1 facet joint and of the right transverse process at the same level. A consequent magnetic resonance imaging (MRI) revealed a well-defined, bilobular subdural extramedullary mass, with a maximum diameter of 3.8 cm at the vertical axis, compressing the spinal cord (Fig. 1).

He underwent emergency T11-L1 wide laminectomy, combined with facetectomy on the affected levels, exploration of the subdural space, debulking of the tumor and T10-L2 posterolateral transpedicular stabilization and fusion, while an intradural, extramedullary mass, causing severe cord compression, was found. The mass was completely removed through microsurgical dissection from the surrounded nerve roots. The maternal root failed to be identified. Hence, it was assumed that it had been already damaged by the tumor, apparently without significant functional value.

No intraoperative signs of peri-or intra-tumor bleeding were observed. After the tumor’s complete excision, the spinal cord was sufficiently decompressed. The timeframe between the injury and the surgical intervention was approximately 8 h. The excised tumor was stored in 10% buffered formalin and sent for histopathological examination. Routine histopathological analysis by hematoxylin/eosin stain revealed a neuronal sheath tumor which was consist of alternatively by cellular areas with numerous Verocay bodies (Antoni A) and few less cellular areas (Antoni B) without any signs of atypia, necrosis and intratumoral hemorrhage (Fig. 2). The tumor was consistent with schwannoma.

The postoperative period was uneventful with rapid recovery. Both motor and sensory functions improved gradually. At the 4th postoperative day, he had regained muscle strength in both lower limbs (2/5 power). He was discharged and he followed a rehabilitation program strengthening his muscles for several weeks.

Six months later he had regained full strength of both lower limbs, being ambulant without support. He was followed up for a total of 44 months, being in excellent condition, fully active, without neurological deficits. MRI scans at 6 and 44 months after surgery, revealed normal spinal canal, in comparison with the schwannoma occupied canal, revealed in the preoperative MRI scan (Fig. 3).

Discussion

Spinal schwannomas account for one third of the primary spinal neoplasms, while the clinical presentation of these tumors is usually related to their location. They typically present with symptoms and signs of myelopathy or nerve root compression, as well as cauda equine syndrome (3). Symptoms are insidious and slowly progressive, due to the slow tumor growth. Early diagnosis and surgical removal are critical, since long-term spinal cord compression may lead to permanent neurological deficits (4). Thorough physical examination in combination with MRI of the area are highly diagnostic. Acute paraplegia as presenting symptom is extremely rare and almost always associated with acute intratumoral hemorrhage (4).

Table I summarizes the reported cases of spinal schwannomas with intratumoral hemorrhage (traumatic or spontaneous), presented with acute neurologic deficit (3-24). There is a limited number of cases with rapidly progressive neurological deficits due to acute cord compression, caused by tumor bleeding. History of prior symptoms is uncommon (8,13,14,16,20). Sudden cord compression caused by tumor bleeding is similar to spinal shock (20). Nine traumatic cases with intratumoral bleeding have been reported so far (8,13,16,19-24).

Spontaneous intratumoral hemorrhage is not uncommon in nervous system tumors. Two main theories exist regarding the hemorrhage’s etiology. The mechanical theory supports that loading causes traction of the tumor vessels resulting in bleeding, while the vascular theory postulates that hemorrhage is caused by spontaneous thrombosis of the tumor vessels, ischemic necrosis and secondary bleeding (11,20,23,25).

Spinal injury after a traumatic event at the level of the tumor could possibly lead to bleeding, resulting in acute canal stenosis and nerve compression (19). In the present case hemorrhage within the tumor or the spinal canal, although suspected and looked for, was not identified.

Mahadewa et al (3) have also reported a patient with acute onset of paraplegia without evidence of intratumoral bleeding. However, in that case the patient had a history of stable back pain and numbness in both legs for 4 years. MRI had revealed an enhancing extra axial mass in the spinal canal, but the patient had declined surgery. Sudden onset of paraplegia could be possibly due to compression of neural elements due to infarction of a significant artery. However, in cases of neurological recovery without surgical intervention, this explanation cannot be supported (3).

In the present case, a hypothesis about the sudden onset of paraplegia could be made: mechanical loading caused by the fall in an already, due to the tumor, narrow spinal canal, resulted in acute neurological damage. Preexisting canal stenosis has been proven to be independent risk factor for developing spinal cord injury, even with minor trauma and without the presence of fracture (26). Spinal canal stenosis may be the reason for the discrepancy between the significance of the trauma and the severity of its results, since acute spinal cord injury following minor trauma has been
The spinal cord may become more vulnerable against external force when the degree of cord compression exceeds a certain threshold (27).

Little is known regarding the growth pattern of spinal schwannomas. Although radical excision is recommended for symptomatic tumors, the optimal treatment for the asymptomatic ones remains unclear. Radiological features, such as heterogeneous intensity on T2-weighted MRI images, may provide useful information regarding the growth potential of spinal schwannomas. In a retrospective study of 23 patients reported (27).
Table I. Reported cases of spinal schwannomas with intratumoral hemorrhage (traumatic or spontaneous), presenting with acute neurologic deficit.

| Authors, year | Study type | Presentation | Spine region | Trauma | Prior symptoms | Treatment | Histology | (Refs.) |
|---------------|------------|--------------|--------------|--------|----------------|-----------|-----------|---------|
| Smith, 1985   | Case report| Cervical     | Cervical     | No     | Severe left scapular pain for 2 weeks (night pain) | Surgical  | Schwannoma intratumoral hemorrhage | (5)     |
| Lee and Lui, 1992 | 2 cases   | Paraplegia   | Thoracic and thoracolumbar | No     | i) 10-month history of back pain ii) 2-year history of low back pain | Surgical  | Neurofibroma with focal haemorrhage | (6)     |
| Uemura et al, 1998 | Case report | Paraparesis | Thoracic     | No     | Occasionally slight sharp pain in right thigh | Surgical  | Schwannoma intratumoral hemorrhage | (7)     |
| Cohen et al, 2000 | Case report | Paraplegia   | Thoracic     | Minor injury (fall from a ladder) | None | Surgical | Schwannoma intratumoral hemorrhage | (8)     |
| Ng, 2001      | Case report | Left hemiparesis | Cervical | No     | 3-day history of sudden onset of left shoulder pain radiating to the left forearm and hand | Surgical  | Schwannoma intratumoral hemorrhage | (9)     |
| Tanaka et al, 2002 | Case report | Paraparesis  | Thoracic | No     | 3-year history of intermittent episodes of lower back pain | Surgical  | Schwannoma intradural hemorrhage | (10)    |
| Parmar et al, 2004 | Case report | Intracranial subarachnoid hemorrhage | Thoraco-lumbar junction | No | History of vague generalized backache for many years | Surgical | Schwannoma intratumoral hemorrhage | (11)    |
| Mahadewa et al, 2005 | Case report | Paraplegia   | Lumbar | No | Stable back pain and a 4-year history of lower-extremity numbness bilaterally 3-day history of neck pain | Surgical | Schwannoma intratumoral hemorrhage | (3)     |
| Ciappetta et al, 2008 | Case report | Myelopathy | Craniocervical junction | No | 3-day history of neck pain | Surgical | Schwannoma intratumoral hemorrhage | (12)    |
| Sharifi et al, 2009 | Case report | Severe neurological deficit | Thoracolumbar (motor-vehicle accident) | Motor-vehicle accident | None | Surgical | Multiple schwannomas | (13) |
| Yeh et al, 2011 | Case report | Paraplegia   | Thoracic | No | None | Surgical | Schwannoma intra-tumoral hematoma | (14)    |
| Kukreja et al, 2014 | Case report | Intracranial subarachnoid hemorrhage | Cauda equina | No | 3-month history of seizures A few days history of left leg pain | Surgical | Schwannoma intratumoral hemorrhage | (15)    |
| Jenkins et al, 2015 | Case report | Paraplegia   | Thoracic | Minor (torsion) | None | Surgical | Schwannoma, intratumoral hemorrhage | (16)    |
| Sahoo et al, 2015 | Case report | Quadripareisis | Cervical | No | Weakness in both upper and lower limbs for 1 day and neck pain radiating to shoulder for 2 days | Surgical | Schwannoma intra-tumoral hematoma | (17)    |
with 5 years mean follow-up, the authors reported absolute relative tumours' growth rates of 139 mm³ and 5.3% per year, respectively. Homogeneously hyperintense or heterogeneously intense on T2-weighted images tumors were significantly larger than the isointense ones at the initial examination. Tumors isointense on T2-weighted images increased very little in volume. However, the schwannomas heterogeneously intense on T2-weighted images had a significantly greater absolute growth rate (28). Most asymptomatic schwannomas have only minimal growth and do not need surgical excision. Close clinical and imaging monitoring is required when patients have large-volume tumors that are heterogeneously intense on T2-weighted images. Surgical removal should be considered in cases of constant tumor growth with significant compression of the spinal cord and cauda equina (28).

The present patient retained normal spinal alignment during the follow-up. Spinal deformities arise in up to 18% of adults and 100% of children after laminectomy for spinal cord tumor excision. Although the necessity of fixation in children has been already documented, evidence supporting concomitant fusion after spinal cord tumors removal in adults is limited (29).

Kobayashi et al (30) recently examined the records for 32 adults who underwent excision of thoracic spinal cord tumors by multilevel laminectomies without fixation. They concluded that even without fixation, sagittal alignment remained unchanged following tumors' excision in the middle and lower thoracic spine, suggesting that fixation may not be necessary. On the contrary, when the tumor is located at the upper thoracic spine, postoperative kyphosis may increase. However, this study had several limitations including the retrospective study design, the small sample size and the lack of a control group treated by laminectomy for a different thoracic spinal disorder (30).

Avila et al (29) reviewed the criteria for fusion following spinal cord tumor resection in adults. The main criteria for fusion were: Preoperative deformity, three or more levels of laminectomy, laminectomy encompassing a spinal junction, young age, facetectomy >50%, persistent deformity 1 year after surgery and C2 laminectomy. Based on these data, in the present case, posterior transperpendicular fixation following tumor removal was performed.

Taking into account the existing literature, this is the first case describing a spinal intradural schwannoma causing acute paraplegia in a previously asymptomatic patient in the setting of trauma, without evidence of bleeding. Acute neurological deterioration, such as paraplegia, may not only be the result of intratumoral hemorrhage but may also occur as a post-traumatic event, possibly due to violent movement or edema formation within a marginally narrowed spinal canal. This possibility should be seriously considered in the management of patients with such tumors and represents additional reason for early surgery. Furthermore, in cases of acute post-traumatic neurological deficits that are not sufficiently justified by the injury per se, the presence of intracanal tumor should be included in the differential diagnosis. MRI remains the main diagnostic imaging technique in such cases. Urgent canal decompression and surgical excision of the tumor is the treatment of choice.
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Availability of data and materials

All data generated or analyzed during this study are included in this published article.

Authors’ contributions

DAK, CT and CK made substantial contributions to the conception and design of the current study. VC, AV, GS and KA acquired and analyzed the data. DAK, CT and CK drafted the manuscript. VC, AV, GS and KA critically revised the manuscript. KA and CK confirm the authenticity of all the raw data. All authors read and approved the final manuscript.

Ethics approval and consent to participate

Not applicable.

Patient consent for publication

Written informed consent for publication was received from the patient.

Competing interests

The authors declare that they have no competing interests.

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