A Long Segmental Hemorrhagic Spinal Schwannoma with Atypical Presentation

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ABSTRACT

Acute hemorrhagic presentation of spinal schwannoma is an event rarely seen. We had ever reported a 63-year-old male presenting with acute spinal cord compression caused by a long segment of intradural extramedullary schwannoma with hemorrhage and atypical enhancement pattern. The magnetic resonance image findings included a long segmental irregular contour, intermediate signal intensity on T1-weighted images, heterogeneous and foci of low signal intensity on T2-weighted images and segmental enhancement of the lesion, all of which is highly atypical for schwannoma. Emergency surgical decompression was performed and the pathologic results have proved the existence of spinal schwannoma.

CASE REPORT

This 63-year-old male patient has presented with progressive bilateral lower leg weakness for 3-4 days, which is associated with dysuria and urinary incontinence. He was brought to our emergency department with sudden onset of paraplegia. He denied any history of trauma, diabetes mellitus, renal disease or other systemic disorders. Physical examination showed positive low back knocking pain. No kyphosis and scoliosis. Pulses in bilateral dorsalis pedis arteries were present. Sensation was markedly decreased to light touch and pinprick below the level of thoracic vertebra T9 dermatomes.

Neurological examination revealed full muscle power in upper extremities and absent in lower extremities. Deep tendon reflexes were absent in lower extremities. No Babinski's sign bilaterally, the initial impression of paraplegia is due to acute spinal cord compression.

Emergency magnetic resonance imaging (MRI) of the thoracic and lumbar spine revealed a long segmental irregular contour of intradural extramedullary lesion which extended from thoracic vertebra T7 to T12 level. The lesion showed intermediate signal intensity on T1-weighted images (Fig. 1a) and heterogeneous signal intensity and foci of low signal intensities within the lesion on T2-weighted

Spinal schwannomas are benign tumors arising from spinal nerve root sheaths [1]. Intraparaspinal nerve sheath tumors are generally slow growing and are diagnosed after causing symptoms such as back pain and progressive neurological symptoms [2]. These intradural extramedullary tumors are most common during the fourth to the sixth decade of life [3]. They rarely present with acute emergency condition. We reported a rare case of long segmental spinal schwannoma presenting clinically with an acute spinal cord compression caused by hemorrhage, which illustrates the diagnostic difficulties for neuroradiologist and neurosurgeons.
images (Fig. 1b, 1c). On gadolinium-enhanced T1-weighted images showed lobulated enhancement pattern (Fig. 1d, 1e). So, the initial impression was ependymoma with spinal cord compression. Other differential diagnosis was astrocytoma, meningioma and hemangioblastoma.

Then the patient was admitted to surgical intensive care units and the operation was done for emergency decompression of the spinal cord. Laminctomy was performed from thoracic vertebrae T7 to T12 level with subtotal excision of the intradural extramedullary tumor. Intra-operatively, the tumor was found as yellowish, easy bleeding with a thin wall capsule and intra-tumoral hematoma.

Grossly, the specimen consisted of multiple pieces of gray white soft tumors measuring up to $1.8 \times 1 \times 0.5$ cm. Microscopic findings showed a picture of schwannoma, composed of Antoni A areas and Antoni B areas (Fig. 2). Hyalinosis of the vessels and Verocay’s bodies were noted.

Reoperation was done due to residual tumor being detected in the follow up MRI. After the operation, the neurological condition was not recovered. Paraplegia with myoclonus on left leg was found. So, he was consulted for rehabilitation. MRI obtained 6 months after surgery, revealed no residual or recurrent tumor.

**Figure 1.**

- **a.** MR imaging of cervical, thoracic and lumbar spine showed long segmental intraspinal lesion from T7 to T12 with cord compression and intradural extramedullary heterogeneous lesion on sagittal T1 FLAIR with some bright signal intensities which showed hemorrhagic areas (arrows).
- **b.** Sagittal T2 showed heterogeneous signal intensity with foci of low signal intensity correlate with hemosiderin depositions (arrows). The spinal cord is compressed to the left side which showed cerebrospinal fluid (CSF) cleft adjacent to the upper and lower boundaries of the tumor (thick short arrow).
- **c.** Axial T2 showed spinal cord (thick short arrow) is compressed to the left side by the tumor (arrow) containing hemosiderin deposit.
- **d,e.** Sagittal and axial T1 fat suppression with contrast enhancement shows segmental enhancement of the lobulated lesion (arrows).
- **f.** Myelography showed the spinal cord is compressed with CSF cleft adjacent to the upper and lower boundaries of the tumor (thick short arrow).
Atypical hemorrhagic spinal schwannoma

DISCUSSION

Spinal schwannomas (neurilemmomas or neurinomas), together with neurofibromas, constitute common intraspinal tumors, representing 25% to 30% of all intraspinal masses [4]. Schwannomas are typically a round or oval, lobulated, and encapsulated tumors arising eccentrically from the nerve root sheath. Schwannomas are intradural in 70% to 75%, extradural in 15%, combined intradural and extradural (dumbbell tumors) in 15%, and intramedullary in less than 1% of patients [4]. They tend to extend through the dural root sleeve into the intervertebral foramen to form a dumbbell-shaped appearance [5].

MRI characteristics of spinal schwannomas are a typical hypointense in relation to the spinal cord on T1-weighted images and hyperintense on T2-weighted images [6]. They may be inhomogeneous on T2-weighted images with focal areas of hyperintensity and hypointensity corresponding to cyst formation, hemorrhage, dense cellularity, or collagen deposition [7]. Virtually all schwannomas are enhanced after administration of contrast medium [4]. Patchy enhancement has been reported in tumors with cystic and/or necrotic components [8].

At present, MR imaging is definitely a choice method for the diagnosis of spinal cord tumors; however, a definitive diagnosis based on signal intensity and enhancement pattern remains difficult to make a differentiation from other spinal tumors [9]. Intramedullary spinal tumors are astrocytoma, ependymoma and hemangioblastoma. Astrocytoma and ependymoma are most commonly seen. They are common in thoraco-lumbar region [9]. Intradural extramedullary spinal tumors are meningioma, lipoma, arachnoid cyst and inflammatory granuloma [9].

MR imaging of ependymomas and astrocytomas shows iso- or hypointense on T1-weighted images and hyperintense on T2-weighted images. After contrast medium enhancement, ependymomas and meningioma reveal intense homogeneous enhancement of the mass. But cystic and hemorrhagic foci are more common in ependymomas [4]. In glioma, enhancement would vary according to the tumor grade; as the low grade gliomas would not enhance [10].

In our case, the schwannoma displayed on an atypical MRI findings with intermediate signal intensity on T1-weighted images and some bright signal intensities which showed hemorrhagic areas (Fig. 1a), heterogeneous signal intensity with foci of low signal intensity correlate with hemosiderin depositions (Fig. 1b). So our first impression was ependymoma. When the pathology report showed intradural extramedullary spinal schwannoma, then we reviewed our case. We recognised the tumor compressed the spinal cord to the left side which showed cerebrospinal fluid (CSF) cleft adjacent to the upper and lower boundaries of the tumor (Fig. 1b, 1f). So, this is a case of a long segmental intradural extramedullary spinal schwannoma with atypical MRI enhancement pattern.

Delayed clinical presentation is common in spinal tumors because these tumors are growing slowly, and the surrounding anatomic environment is permissive while the presenting symptoms are always non-specific [11].

Hemorrhage is unusual in spinal schwannoma but

Figure 2

Immunostaining (X400) positive staining of S-100 protein in the tumor nuclei, composed of Antoni A areas and Antoni B areas (arrows).
more common finding in ependymoma. The exact mechanism of initiation of hemorrhage in schwannomas remains obscure. One theory proposed that the hyalinized ectatic vessels seen in schwannoma undergo spontaneous thrombosis. This is followed by distal tumor necrosis that may be complicated by hemorrhage. Another theory postulates that hemorrhage occurs as a result of traction on the tumor vasculature during movements [12]. The latter theory may explain the association with physical stress in some cases that tumors are located in the mobile segments of the spine.

In our case, microscopic finding shows hyalinosis of vessels. So the former may be more appropriate for this case [12]. Complete excision of tumor may prevent recurrence. To our patient, complete excision was successful but due to extensive tumor involvement, neurological condition cannot recover. A successful surgical outcome depends on early diagnosis and complete surgical excision [11].

CONCLUSION

In conclusion, the case we have reported of intradural extramedullary schwannoma with longitudinal extension in the thoracic region is very rare and has never been reported in the past because schwannoma usually extends into the enlarged neural foramen forming a dumbbell shaped paraspin mass. Atypical MR findings illustrated the diagnostic difficulties. So, schwannoma should be considered as the differential diagnosis of an intradural extramedullary tumor with spontaneous hemorrhage.

REFERENCES

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