CASE REPORT

Ancient Schwannoma of the Conus Medullaris

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SUMMARY

Cystic spinal or ancient schwannoma is a rare form of intradural tumour especially in the conus medullaris region. Due to its indolent behavior and benign course, the diagnosis of schwannoma is always a challenge and the imaging findings can be misleading. Our patient presented with chronic mild sciatica pain without any other neurological symptom. MRI of the spine showed intradural tumour located at the conus medullaris region with nerves compression and was reported as ependymoma. L3-L5 laminectomy and total excision of tumour was performed without any neurological complication. We concluded that ancient schwannoma of the conus medullaris is a rare benign tumour that can present with minimal non-specific neurological symptom that lead to misdiagnosis. Radical tumour excision is safe with an excellent outcome.

KEY WORDS:
Ancient schwannoma, Sciatica, Radical excision

CASE REPORT

We report a case of 54 years old Malay man presented with history of progressive low back pain over a period of two years. The pain was described as dull in nature, occurring intermittently and maximum on bending or flexing the right thigh. It was gradually getting worse in the last three months with radiation to the right leg. He had history of fall few years before the onset of back pain. There was no other associated neurological symptom such as limb numbness or weakness. He denied any history of bowel or urinary disturbance. Examination of the neurological system revealed a positive straight leg raising (SLR) test of the right side. The power of both leg muscles were normal with no features of pyrimidal tract sign. There was no sensory lost and anal tone was also normal. He was diagnosed as prolapsed lumbar disc with radiculopathy and subsequently referred to us for further management.

A Magnetic Resonance Imaging (MRI) of the lumbosacral region revealed a mass lesion in the conus medullaris region with spinal nerves compression. The lesion was located intradurally and measured 7.1cm in length. It enhanced heterogeneously following contrast administration (Figure 1). The lumbar vertebras were normal and there was no disc prolapse seen. It was reported by the radiologist as conus medullaris ependymoma.

Patient underwent L3-L5 laminectomy and excision of the tumour. Intra-operatively we found a well rounded lesion in

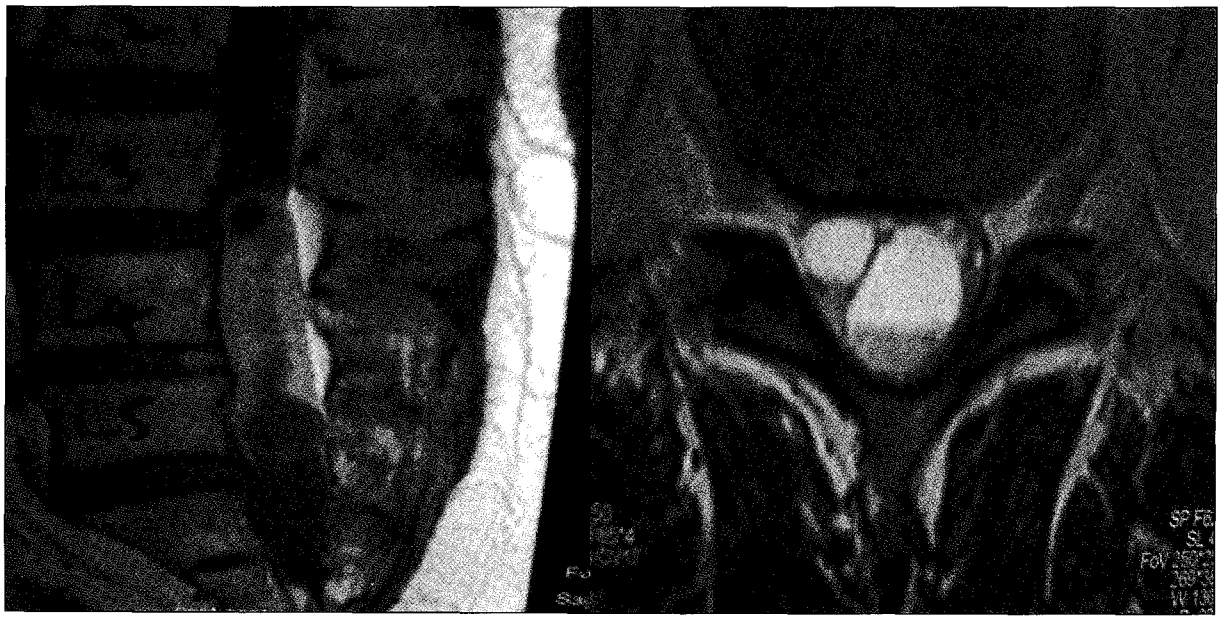

Fig. 1: MRI showing an intradural lesion at L3-L5 vertebra level which was heterogenously enhanced with contrast (A). On axial view of T2WI, this lesion appears cystic in nature as evidenced by multiple fluid levels (B). The spinal nerves are displaced laterally.

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Fig. 2: Intra-operative photo showing tumour in the conus medullaris region attached to the sacral nerves.

the conus medullaris region. The tumour was cystic in nature and firm in consistency. It had grayish fluid in the centre and the ventral part of the tumour was attached firmly to a spinal nerve possibly L4 (Figure 2). Using the microscope with the aid of SSEP we managed to excise the tumour in total without causing any nerve injury. There was no much of bleeding and the surgery went smoothly. The spinal nerves were left freely intact in a large CSF cavity upon closing the dura mater. Lower limb SSEP showed normal wave and amplitude throughout the surgery.

Patient showed excellent recovery following surgery. His back pain had completely disappeared. He had no neurological deficit after the operation. The wound healed well with no sign of CSF leakage. He was discharged home after sutures were opened. The specimen that was sent came back with a surprising report. Microscopic H&E staining of the tumour showed typical features of schwannoma. The cells were spindle in shape forming small areas of compact (Antoni A) and loose cellularity (Antoni B). There were few cystic spaces seen. Some cells were pleomorphic with no mitotic activity seen (Figure 3). The final histopathology diagnosis was spinal cystic schwannoma (Ancient schwannoma).

DISCUSSION

Schwannoma is a benign tumour of the peripheral nervous system, arising from Schwann cells. Schwannoma can occur virtually in any body location where peripheral or cranial nerves are present, and they correspond to 8% of primary intracranial and 29% of primary spinal tumors[1]. Prevedello et al. have investigated the prevalence of different extramedullary intradural spinal tumours and observed that schwannoma comprised 66% of all cases[2]. The lumbar region is one of the most common sites for occurrence of spinal schwannoma, as demonstrated by Conti *et al.*, who studied the distribution of spinal schwannomas and reported that 48% lie within the lumbar spine[3].

Schwannoma is a slow growing tumour. It usually encapsulated, and rarely undergone malignant transformation. Schwannoma arises from the Schwann cells of the nerve sheath, and they comprise the most common tumour type affecting the peripheral nerves[1]. The most common location of spinal schwannoma is around peripheral nerves in the extradural space. Intracranial schwannoma have also been observed, and it usually arises from the facial, trigeminal, or vestibular nerves[4]. Women and men are equally affected by schwannoma and there is a predilection for occurrence in between the fourth to sixth decades of life[2]. Benign schwannoma can occasionally display degenerative changes that are encompassed by cyst formation, calcifications, hemorrhage and hyalinization. When multiple degenerative changes are encountered, schwannoma fits into the category of "ancient schwannoma", which is extremely benign in course, rarely demanding any form of treatment[1]. Cystic degenerations have been observed in the orbital region, in the olfactory groove, in the tentorial hiatus and posterior cavernous sinus, in the pre-sacral region, within the pancreas, in the maxillary sinus, within the spinal cord, and intraventricular. Cystic schwannoma have also been observed surrounding cranial nerves such as the vestibular nerve, the vagus nerve and within the jugular foramen[5]. Cystic schwannoma occurring in the conus medullaris region

Fig. 3: Microscopic appearance of the tumour showing two distinctive features of Antoni A (black arrow) and Antoni B (white arrow) areas in Schwannoma. No areas of necrosis or mitosis seen.

is rare. Only seven cases of cystic lumbar nerve sheath tumours have been described in the literature pointing out its resonance magnetic imaging presentation[1].

Ancient schwannoma is a benign tumour with a slow growth rate and the symptoms are very few and minimal. Because of that, the diagnosis of ancient schwannoma is always a challenge to the neurosurgeon and neurologist. In this case, there was an obvious contrast with the paucity of symptom and the size of the tumour. Patient presented with long standing history of sciatica pain without any other neurological symptom such as limb weakness or numbness and incontinence. Examination revealed mild lumbar tenderness without other features of cord or spinal nerve compression. This atypical presentation was mimicking the diagnosis of lumbar disc prolapsed with radiculopathy. Scenario of this case highlights the importance of suspecting an ancient schwannoma in patient with progressive symptoms of disease associated to lumbar spinal cord or nerve roots compression. Moreover, ancient schwannoma should be included in the differential diagnosis of a cystic mass in the spinal region.

Ancient schwannoma or cystic spinal schwannoma is possibly behaves in a similar fashion to solid schwannoma. Therefore, the treatment of an ancient schwannoma is similar to other spinal schwannomas which involve radical surgical excision of the tumour and decompression of spinal nerves[1]. In our case, were performed a wide lumbar laminectomy (L3-L5) and total excision of the tumour. Using the microscope with the aid of intra-operative monitoring (SSEP), we managed to completely remove the tumour that was attached to the spinal nerves in the conus medullaris without any complication. Post-operatively the sciatica pain resolved and patient was discharged home well.

CONCLUSION

In conclusion, ancient schwannoma of the conus medullaris region is a rare benign tumour that grows very slowly. Clinical symptoms are usually due to the compression of lumbar spinal nerve roots structures. Wide CSF space and absent of spinal cord below the L3 level are also factors for its minimal sign even though the tumour size is big. The treatment for ancient schwannoma of the conus medullaris region is radical excision of the lesion and the outcome is always excellent with the availability of modern technology.

REFERENCES

1. Borges G, Bonilha L, Proa M Jr. Fernandes YB, Ramina R, Zanardi V, Menezes JR. Imaging features and treatment of an intradural lumbar cystic schwannoma. Arq Neuropsiquiatr, 2005; 63(3A): 681-4.
2. Prevedello DM, Koerbel A, Tatsui CE, *et al.* Prognostic factors in the treatment of the intradural extramedullary tumors: a study of 44 cases. Arq Neuropsiquiatr, 2003; 61(2A): 241-7.
3. Conti P, Pansini G, Mouchaty H, Capuano C, Conti R. Spinal neurinomas: retrospective analysis and long-term outcome of 179 consecutively operated cases and review of the literature. Surg Neurol, 2004; 61(1): 34-43; discussion 44.
4. Burchardt D, Intraspinal neurinoma: spinal tumor diagnosis in computerized tomography. Aktuelle Radiol, 1997; 7(3): 152-3.
5. Shenoy SN and A Raja, Cystic cervical intramedullary schwannoma with syringomyelia. Neurol India, 2005; 53(2): 224-5.