Spinal Cystic Schwannoma: An MRI Evaluation

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ABSTRACT

Spinal cystic schwannomas are a very rare entity and have been reported in only a few case reports in literature; its diagnosis and management has remained a challenge. This study reviewed the results of magnetic resonance imaging (MRI) of 12 patients (7 men and 5 women; aged 37 - 67 years; mean age: 52.75 years) with pathologically proven cystic schwannoma of the spine and discussed their differential diagnosis. All patients underwent surgery at our institutions between June 2000 and April 2012. MRI showed well-delineated intradural and extramedullary lesion of iso- to low signal intensity on T1 weighted images, high signal intensity on T2 weighted images, and rim enhancement on contrastenhanced images. A precise understanding of the MRI features of spinal schwannomas, especially the typical characteristic of enhancement, may help clinicians in their pre-operative diagnoses and surgical planning.

Key Words: Spine. Cystic. Schwannoma. MRI. Contrast enhancement.

Spinal schwannomas are frequently observed in neurosurgical practice. However, cystic spinal schwannomas are very rare.1 Because of their indolent behaviour and benign course, the diagnosis of schwannomas may be challenging, and the imaging findings can often be misleading.² Malignant cases have been reported in association with Von Recklinghausen's disease. Schwannomas can show a variety of degenerative changes such as cyst formation, calcification, haemorrhage and hyalinization.² Cyst formation usually results from the degeneration of the Antoni B portion of the schwannoma and central ischaemic necrosis by tumour growth.3,4 In this report, the aim was to describe the clinical presentation and magnetic resonance imaging (MRI) characteristics of spinal schwannomas and elucidate the importance of contrast-enhanced MRI in their differential diagnosis.

The magnetic resonance (MR) images of 12 patients (7 men and 5 women; age range: 37-67 years; mean age: 52.75 years) with pathologically proven cystic schwannoma of the spine were reviewed. All patients were treated surgically at our institution between June 2000 and April 2012 and followed-up for 6 months to 2 years. Patient information regarding clinical presentation, tumour location, and operative findings was collected retrospectively from medical records. All MR images were obtained using a Philips Gyroscan Intera 1.5T scanner with a spine surface coil. The signal intensity of the lesions was compared to adjacent normal tissue in T1 weighted images (T1WI), T2 weighted

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images (T2WI), and contrast-enhanced images. MRI of all patients showed well-delineated intradural and extramedullary (IDEM) lesions of iso- to low signal intensity on T1WI, high signal intensity on T2WI, and strong ring-like enhancement of the cyst wall on contrast-enhanced images (Figures 1 and 2). Three patients showed a lesion in the cervical region (25%) with no sign of any bony erosion and scalloping; one patient showed a lesion in the thoracolumbar region, extending from 6th thoracic (T6) to 1st lumbar vertebra (L1) (8.33%), with bony scalloping on T10 to T12 vertebrae, yet no sign of bony erosion. Spinal cord was compressed posteriorly with a marked foraminal widening.

Eight patients had lesions in the lumbar region (66.66%), five showed bony scalloping but no sign of bony erosion, and two had slight bone erosion in the lumbar L4 and L5 vertebrae. Bony erosion was less frequent in our cases, which may be attributable to the cystic nature of the lesions. All lesions were cystic because of their mass effect, causing symptoms and intraoperative identification of the cystic element, as well as positive MRI findings. No remarkable septation within the cyst was observed. The lesions involved 1-8 vertebral segments, with an average of 2.5 vertebral segments. The average duration of the symptoms ranged from 2-60 months (mean duration: 17.66 months).

The most common initial symptom was radicular pain followed by motor weakness. Myelopathy, non-specific back pain, long-term history of vague flank pain, bladder/bowel incontinence, sciatica, and radiating pain in the bilateral lower limbs were also reported. All these symptoms were frequently found in patients with lesions in the lumbar spine. Patients with lesions in the cervical spine reported progressive neck pain in the upper cervical region, paraparesis over a period, radiating pain, feeling of discomfort, and myelopathy. The patient with the largest lesion in the thoracolumbar spine,



Figure 1: Median sagittal T1WI (1A) and T2WI (1B) of cervical spine (C5 to C7) shows a well defined IDEM lesion, isointense to CSF on T1WI and T2WI (1A, thick arrow and 1B, thin arrow). A thick ring-like enhancement of the lesion with irregular appearance is observed during contrast T1WI (1C, dotted arrow).



Figure 2: Median sagittal T1WI (2A) and T2WI (2B) of the lumbosacral spine showing an intrathecal lesion at the L1 level (2A, thick arrow and 2B, thin arrow). A ring-like enhancement of the lesion is observed during contrast T1WI (2C, dotted arrow). A sacral meningeal cyst was incidentally discovered at S2 and shows no enhancement (2C, arrow head).

extending from T6 to L1 vertebral segments, had paraparesis, back pain, numbness in both legs, and right radiculopathy. Postoperatively, all patients achieved complete relief and did not experience any of the previous reported symptoms. No recurrent tumour was found during the follow-up period of 6 months to 2 years.

Schwannomas are slow-growing, mostly encapsulated, and solid benign tumours. The most common location of schwannomas is on the peripheral nerves or spinal nerve roots in the extradural space. The tumour commonly arises from the dorsal sensory roots of the cervical and lumbar spine with less frequent involvement of the thoracic region.^{2,5,6} This study showed that a higher frequency of schwannomas in the lumbar spine (66.66%). Men and women are equally affected and may become symptomatic in patients at any age, but its peak incidence is around the fourth to sixth decades of life.3,5 In this study, the mean age was 52.75 years and peak incidence was also between the 4th and 6th decades. Cystic tumours have a high risk of causing progressive symptomatic worsening as a result of cyst expansion. Because of its slow-growing nature, few symptoms can be observed until the tumour has reached a large mass. Histologically, schwannomas consist of compact cellular lesions (Antoni type A) and loose, hypocellular myxoid

lesions with microcystic spaces (Antoni type B). Immunohistochemically, these tumours show diffuse positivity for the S100 protein in the cytoplasm of the tumour cells.

MRI is the choice of modality for the evaluation of intradural spinal tumours. Schwannomas tend to have signal intensity equal to or less than that of the spinal cord on T1WI and mild to marked hyperintensity on T2WI. Schwannomas usually show fluid signal intensity on T2WI, which could be a useful predictor in its diagnosis. Bony scalloping and bony erosion usually occur in schwannomas. Enhancement is variable and can be intense and homogeneous in some lesions, while it may only show peripheral enhancement in other cases. A contrast study is necessary to differentiate these from other cystic neoplasms. Schwannomas enhance very strongly, most of the time with an irregular appearance. A cystic schwannoma has a thick wellenhancing rim enhancement of the cyst wall compared to other cystic lesions. Strong rim enhancement of an IDEM tumour during MRI should be considered in the diagnosis of schwannomas, 1,2,6 which was evident in all these cases. Nevertheless, the accurate diagnosis should be proven by pathological studies.

The differential diagnosis of schwannomas is primarily based on location, clinical presentation, age, and gender of the patient. The differential diagnoses of intradural extramedullary cystic schwannoma include cystic meningioma, arachnoid cyst or perineural cyst, epidermoid cyst, dermoid cyst or cystic teratoma, neuroenteric cyst, hydatid cyst, and cystic ependymoma. Cystic meningiomas are found more typically adjacent to the tumour and less commonly within the tumour. Women are frequently affected, with a high incidence in middleaged patients, and majority of cases are found in the thoracic region. The incidence of osseous erosion and soft-tissue calcification is frequently observed. After contrast administration, a thin region of enhancement contiguous with the dura (dural tail sign) is often observed. In addition, cysts within the tumour are usually eccentric, and ring-like enhancement is rarely reported. Arachnoid cysts are isointense to cerebrospinal fluid (CSF) on all pulse sequences and do not show enhancement. They are usually located posterior to the thecal sac and primarily occur in adolescents and young adults. Epidermoid cysts are slightly hyperintense to surrounding tissue on T1WI and show similar intensity to CSF on T2W images. They have lobulated margins with peripheral enhancement or no enhancement during contrast-enhanced MRI. The cystic lesion of epidermoids generally shows low signal intensity on the TIWI relative to the nerve tissue. Dermoid cysts are generally seen with a dermal sinus tract and show high signal intensity that is suggestive of fat tissue. Neuroenteric cysts are multi-lobulated, extended, and are usually located in the anterior intradural extramedullary

region. Hydatid cysts have multiple well-circumscribed cystic lesions, internal echoes, and daughter cysts. Ependymomas are solid in nature, although mucinous degeneration may occur. The mucinous material of proteinaceous nature increases the signal intensity in the cystic ependymoma on T1WI. The intramedullary location and / or connection to the conus medullaris or filum terminale is helpful for diagnosing cystic ependymomas. 1,5,6

In conclusion, MRI is the modality of choice in the evaluation of spinal cystic schwannomas. Careful analysis of the patient's clinical findings and an accurate understanding of the MRI features such as age, gender, tumour locations, widening of the neural foramen, fluid signal intensity on T2WI, strong ring-like enhancement, or rim enhancement patterns of the cyst wall without a dural tail sign may assist clinicians in their pre-operative diagnoses, guide the surgical procedure, and facilitate in their differential diagnosis.

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