Case Report

Multiple spinal ring-enhancing schwannomas

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ABSTRACT

We present a case of intradural multiple ring-enhancing lesions in a 20-year-old male with symptoms and signs of progressive spastic paraparesis. An MRI of the thoracolumbar area showed 2 peculiar ring-enhancing lesions, at the level of T12 and L1. The differential diagnosis includes inflammatory or infectious lesions in addition to rare cystic tumors. The patient underwent surgical resection of the 2 lesions with an uneventful perioperative course. Histologically, the diagnosis was consistent with cystic schwannomas.

Neurosciences 2005; Vol. 10 (1): 101-102

Nerve sheath tumors like schwannomas or neurofibromas are one of the most common intradural-extramedullary tumors of the spine.[1] They are commonly found in neurofibromatosis type I patients. However, cystic degeneration of schwannomas is extremely rare.[2] Intradural spinal ring-enhancing lesions have a differential diagnosis including infectious processes and cystic enhancing tumors. This case report describes the clinical presentation, neurological images, and clinical course of a patient with multiple intradural-extramedullary ring-enhancing schwannomas.

Case Reports. A 20-year-old male with no cutaneous stigmata of neurofibromatosis type I and a non-contributory family history presented with a 4-month history of progressive paraparesis and paresthesia of the lower-extremities. He denied any bowel or bladder symptoms. On examination, the muscle tones of lower-extremities were normal. Motor power was $4^{+}/5$, deep tendon reflexes were slightly exaggerated with equivocal plantar responses bilaterally. Plain radiographs of the lumbar spine were normal. An MRI of the thoracolumbar spine revealed 2 intradural-extramedullary lesions (**Figure 1**). The 1st lesion was located opposite the T12 vertebral body and measured about 12 mm. The lesion was isointense on T1-weighted images and hyperintense on T2-weighted images with a strong ring-enhancing pattern after gadolinium-diethylene triamine penta-acetic acid (Gd-DTPA) administration. The first lesion caused lateral displacement of the spinal cord and signal changes within the cord on T2 - weighted images (**Figure 2**). The 2nd lesion was more elongated at the level of the first lumbar vertebra (L1), measured approximately 20 mm with similar signal intensity and ring enhancement after gadolinium injection. The MRI features were more suggestive of inflammatory or infectious process; however, rare cystic intradural tumors are within the differential diagnosis. The patient underwent surgical intervention with T12 and L1 bilateral laminectomies. A gross-total resection of the intradural-extramedullary lesions was achieved. The patient had an uneventful postoperative course with no new neurological deficits. Over a 4-month period, his presenting symptoms started to improve. Histological examination of the lesions revealed well-capsulated spherical lesions with central cystic

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Received 30th May 2004. Accepted for publication in final form 26th July 2004.

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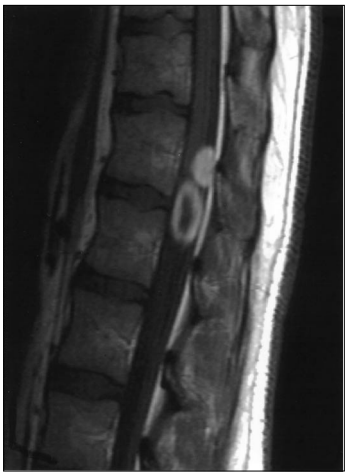

Figure 1 - Sagittal T1-weighted MRI showing 2 lesions, the first at T12 vertebral body measuring 12 mm, and the second at T12-L1 level with ring enhancing pattern (after gadolinium injection).

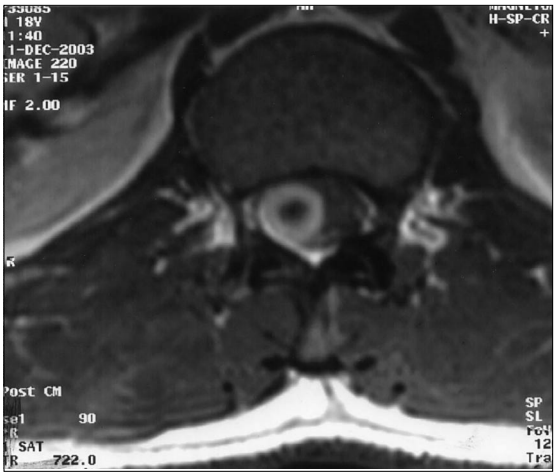

Figure 2 - Axial T1-weighted MRI at the level of T12 vertebral body demonstrating a ring enhancing lesion (after gadolinium injection) causing displacement of the spinal cord.

degeneration. The ring-enhancing tissue was consistent with Antoni type A and Antoni type B areas, with no histological evidence of aggressive changes or increased mitotic figures.

Discussion. The MRI findings of this patient illustrate adjacent ring-enhancing lesions. These lesions are strongly suggestive of inflammatory or infectious pathology similar to an abscess. Benign nerve sheath tumors like schwannomas with cystic-central degeneration are included in the differential diagnosis. However, the pattern of enhancement and the appearance of the lesion wall is not typical of the rarely encountered cystic schwannoma. This case report is interesting in illustrating a rare radiological pattern of cystic schwannomas. This provides a clear example of such benign pathology and how it may mimic a radiologically infectious process like abscesses. Cystic changes may occasionally occur in large schwannomas. Such patterns were reported by Ogose et al,[3] who described a cystic degeneration of presacral pelvic schwannoma (also known as ancient schwannomas). Cystic changes also occur in intradural spinal schwannomas as previously reported by Shen et al[2] and Parmar et al.[1] In both reports they described the enhancing rim of the cystic tumors as irregular thin capsules. In all reported cases of cystic schwannomas in the literature, they were reported as cystic degeneration and not associated with an aggressive clinical course.

A few explanations for the pathophysiology of the cystic degeneration in schwannomas have been proposed. 1. As a result of degenerating Antoni B portion of the tumor, which results in the formation of small cysts. These small cysts can coalesce to form a larger central cyst.[4-6] 2. Insufficient blood supply can result in central ischemia and necrosis secondary to tumor growth.[7] 3. As a hemorrhagic etiology, fluid-levels were identified.[6,8]

In conclusion, cystic schwannomas with ring-enhancing patterns are rare forms of spinal schwannomas and may mimic inflammatory or infectious processes. Such cystic changes are not a sign of more aggressive clinical behavior. Therefore, gross-total resection is advocated as an adequate treatment of choice in these cases.

Acknowledgment. The authors thank Michelle Harper, RN, BSN for help in preparing this manuscript.

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