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CASE REPORT

Redundant Nerve Roots of the Cauda Equina Associated with Lumbar Spinal Arachnoid Cyst

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ABSTRACT

A patient with redundant nerve roots of the cauda equina associated with the presence of an intra-spinal arachnoid cyst is described. She presented with sensory, motor and sphincter dysfunction. Magnetic resonance imaging revealed redundant nerve roots manifesting as serpiginous structures within the thecal sac. Marked improvement of symptoms was noted after laminectomy. The serpiginous appearance of the nerve roots persisted postoperatively despite good decompression of the spine. The clinical and radiological presentation, aetiology, and treatment will be discussed.

Key Words: *Arachnoid cysts; Cauda equina; Magnetic resonance imaging; Peripheral nervous system diseases; Spinal nerve roots*

中文摘要

腰椎蛛網膜囊腫相關的馬尾神經根鬆弛症

陳秀慧、李雷釗、周明德

本文報告一宗與椎管內蛛網膜囊腫相關的馬尾神經根鬆弛症。病人呈感覺、運動及括約肌障礙。磁共振影像顯示神經根鬆弛在硬膜囊內成匐行狀結構。病人接受椎板切除術後，病情明顯好轉。脊柱減壓術成功完成後，病人神經根匐行狀結構依然存在。本文討論有關此症的臨床及放射特徵，以及病因及治療方法。

INTRODUCTION

Redundant nerve roots (RNRs) of the cauda equina are characterised by the presence of tortuous structures in the cauda equina associated with lumbar canal stenosis. The aetiology and the mechanism of the disease remain controversial. Previously, lumbar myelography was used for diagnosis, in which the RNRs manifested as serpiginous filling defects. Resolution of the serpiginous changes after laminectomy has been reported.[1]

We report on a patient with the RNR syndrome associated with an intra-spinal arachnoid cyst diagnosed by magnetic resonance imaging (MRI) of the lumbar spine. Due to the non-invasiveness of MRI, it has mostly replaced myelography as the first diagnostic test in persons presenting with possible spinal pathology.

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The radiological presentation and the clinical aspects are discussed.

CASE REPORT

A 32-year-old woman had low back pain for two years. There was no previous trauma or infection of the spine. She presented with a one-week history of aggravation of the low back pain with associated bladder dysfunction in October 2001. Ultrasonography of the pelvis showed a large amount of residual urine after micturition. There was a change in the bowel habit with reduced number of bowel openings. The patient also noticed paraesthesia over the perineal region and the posterior aspects of both thighs.

Physical examination revealed a reduction in pinprick sensation over the S2 to S5 dermatomes. Plantar flexion of the left ankle was weak. Lower limb deep tendon reflexes and anal tone were preserved, and no other focal neurological deficit was evident.

A plain radiograph of the lumbosacral spine showed loss of the normal lumbar lordosis with reduced disc height at the L4/L5 level. There was no kyphoscoliosis.

Figure 1. (a) A T2-weighted sagittal magnetic resonance image shows a hyperintense cystic lesion dorsal to the cauda equina at L3-L5 level in keeping with intra-spinal arachnoid cyst. In addition there is a loss of bright signal in the L4/L5 disc. (b) A close-up view showing the cauda equina manifests as a cake of serpiginous hypointense filling defects within the thecal sac most evident at L5 level.

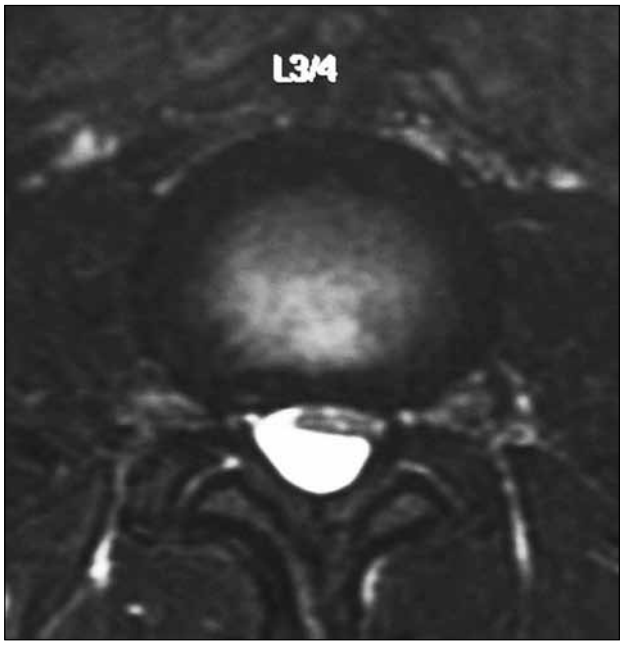

Figure 2. A T2-weighted axial magnetic resonance image shows the cauda equina being compressed and displaced anteriorly by the arachnoid cyst.

Lumbar spine MRI with and without contrast injection was performed using a 1.5 Tesla Signa system (General Electric, Milwaukee, USA). The spinal cord ended at the lower L2 level. A well-defined intra-dural extramedullary thin-walled cystic T2-weighted hyperintense non-enhancing lesion was noted dorsal to the cauda equina at the L3-L5 level (Figure 1a). Mass effect with compression and anterior displacement of the cauda equina were also seen (Figure 2). These findings were suggestive of intra-spinal arachnoid cyst with resultant spinal stenosis. A cake of serpiginous T1-weighted isointense, T2-weighted hypointense lesion was seen extending from mid L5 level to mid S1 level, just below the level of critical compression (Figure 1b). No flow void suggesting an element of high flow was observed, nor was there any abnormal dilated draining vein or leptomeningeal enhancement.

In addition, disc desiccation with associated loss of T2-weighted bright signal was seen in the L4/5 disc, which showed mid-posterior protrusion (Figure 1a).

An L3 to L5 laminectomy was performed. Intra-operatively an intra-dural cystic lesion containing clear cerebrospinal fluid was found. The dura mater was tense and not pulsatile. After fenestration of the cyst, the cauda equina nerve roots were noted to be very elongated and curled up like a bag of worms,

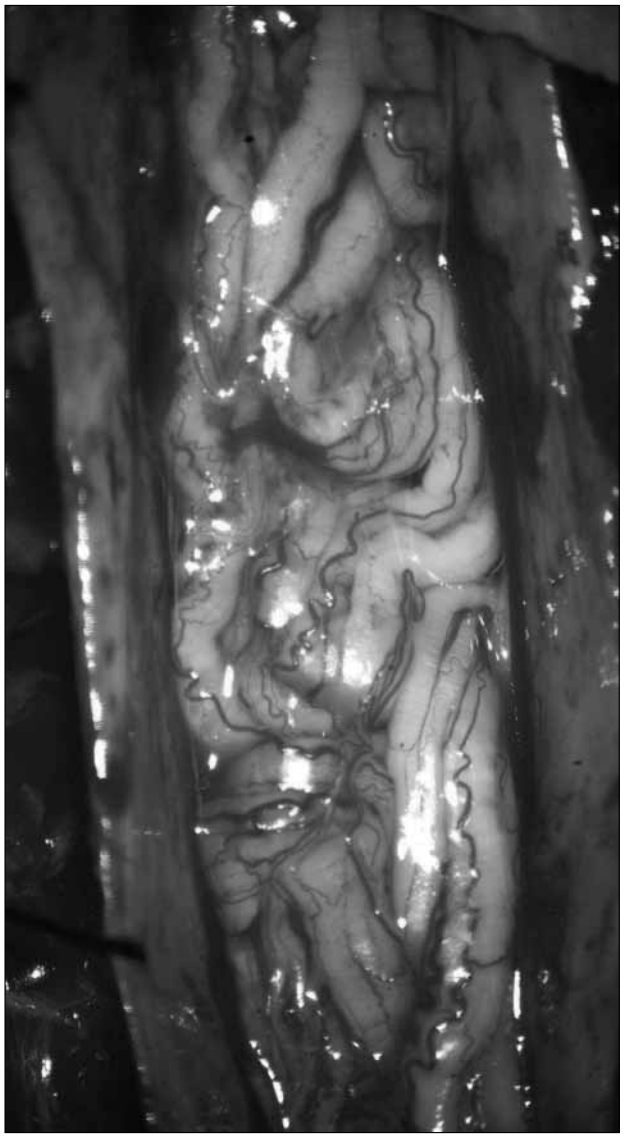

Figure 3. An intra-operative photo showing the presence of tortuous, elongated nerve roots after the dura is opened.

packing themselves at the lower end of the thecal sac. They were not thickened (Figure 3). There was no arachnoiditis. The arachnoid cyst was subtotally excised and its wall fenestrated into the sub-arachnoid space. Histopathological examination of the cyst wall confirmed that it was a meningeal cyst.

Improvement of lower limb numbness and weakness was reported three months after the operation. Bladder and bowel function returned to normal. There was also an improvement in low back pain.

The MRI of the spine was repeated postoperatively at 1 month, 3 months and 32 months after surgery, and showed no evidence of cyst recurrence. The spinal canal

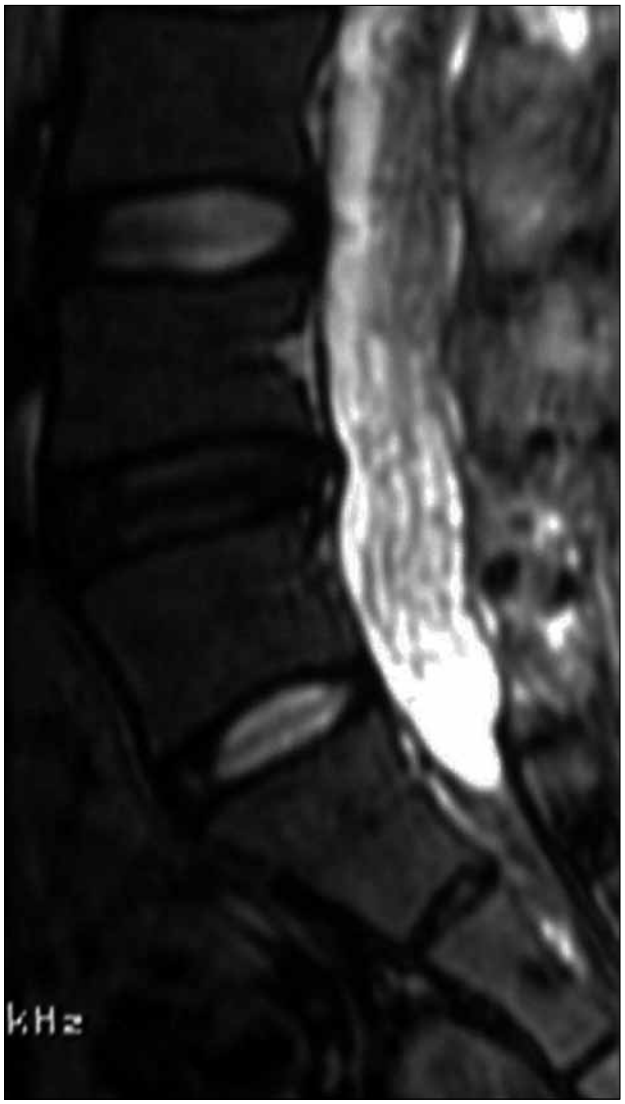

Figure 4. A T2-weighted sagittal magnetic resonance imaging (MRI) scan 32 months after the operation, showing the persistence of serpiginous nerve roots. The cauda equina appears straightened when compared to the preoperative MRI appearance.

and thecal sac were widened and well decompressed. Persistence of the serpiginous nerve roots was still evident on the MRI obtained 32 months after surgery. The roots were surrounded by copious cerebrospinal fluid space, appeared more straightened, and spanned three vertebral bodies (Figure 4).

DISCUSSION

RNRs of the cauda equina are characterised by the presence of elongated and tortuous nerve roots in the subarachnoid space of the lumbar spine. This entity was first described by Verbiest in 1954.[2] There is a male predominance and the age of onset ranges from the fourth to the eighth decade.[3] Most patients present with low back pain and symptoms of intermittent neurogenic claudication, aggravated by exertion and relieved by rest.[1] Bowel and urinary symptoms are relatively less common.[4] However, our patient did not experience any neurogenic claudication. The change in bowel habit and urinary retention, together with aggravation of low back pain, brought her to medical attention.

The aetiology of RNRs remains unclear. It has been suggested that the syndrome is acquired, and most reported cases were associated with spinal canal stenosis secondary to spondylotic changes in the spine.[4,5] Patients with achondroplasia may have RNR,[6] which probably resulted from generalised spinal stenosis associated with the disease. RNRs have also been reported in association with malignant plexiform neurofibroma,[4] as well as with a localised form of hypertrophic neuritis.[7] One previously reported case was associated with the presence of an arachnoid cyst.[8]

The mechanism of RNR formation remains unclear, most have been found above the level of any stenosis, but occurrence below that level have also been reported.[9,10] Earlier studies suggested that their formation was related to the constricting effect exerted by the narrowed segment of the canal, leading to nerve root redundancy above the stenosis and a stretching of nerve roots below it.[4,8,10] However the equal distribution of stretching force upon all the nerve roots could not explain the uneven pattern of their distribution and lengths.[11] It was suggested that a selective force exerted on some nerve roots at the level of the stenosis could account for the pathogenesis of RNR.[11] In situations where redundancy was observed below the level of stenosis as seen in this case, the mechanism of RNR formation is more difficult to explain.[1] In such cases the nerve roots proximal to the stenosis were stretched, producing a redundancy in the distal roots.[5]

In most reported cases of RNRs, lumbar myelography was the most important preoperative diagnostic modality. Serpiginous filling defects, associated with either a partial or complete extradural block and spinal stenosis, were usually evident on the myelogram.[3,5,7] Other differential diagnoses include arteriovenous malformations and venous obstruction from any cause.[1,3] Several features help differentiate RNRs from other differential diagnosis. The disappearance of the serpiginous myelographic filling defects with positioning might be helpful in differentiating RNRs from these other alternatives.[10] In addition,

arteriovenous malformations are associated with large draining vessels that are not seen with RNRs. In patients with engorged veins from any type of compression, they are always below the level of the block.[3] However, serpiginous filling defects are inconsistent myelographic findings in this condition, and the correct diagnosis can only be made with certainty when the dura is opened at surgery.[8]

In a few recent studies, MRI was used as the main diagnostic modality for RNR evaluation,[12-14] as it can demonstrate the presence of RNRs having a similar signal intensity to other cauda equina nerve roots on both T1-weighted and T2-weighted images.[13] It has also been shown that patients with such MRI features generally present with more severe clinical symptoms.[12]

MRI should be the investigation of choice for RNRs, as the level and extent of spinal stenosis can also be reviewed. The RNRs appear as T1-weighted isointense, T2-weighted isointense, or slightly hypointense serpiginous structures within the thecal sac. The lack of flow voids and absence of enlarged draining veins help differentiate the condition from spinal arteriovenous malformations. It has been demonstrated that MRI is as sensitive as myelography for the diagnosis of spinal stenosis, with the additional benefits of being non-invasive and having minimal risk of allergic reactions to contrast.[15]

Most patients experience a marked improvement of symptoms after laminectomy.[1,5,10,11] Complete resolution of the serpiginous change in postoperative myelograms has been observed.[1,11] Although such abnormalities could still be observed on the follow-up MRI 32 months after the decompression operation in our patient, they were less serpiginous and extended over a wider span than in the preoperative MRI. Together with the release of the spinal stenosis, our patient's symptoms showed a significant improvement three months postoperatively. The relief of symptoms after surgical removal of arachnoid cyst and the persistence of serpiginous nerve roots postoperatively suggest that spinal canal stenosis rather than the RNRs caused the clinical manifestations in our patient.

We have reported a patient with RNRs in the cauda equina associated with the presence of an intra-spinal arachnoid cyst. It is important to recognise the MRI features of RNRs to make an accurate diagnosis and exclude other causes of a serpiginous appearance within the spinal canal. If the diagnosis is in doubt, exploration of the spinal canal should be considered. For this condition, patient should receive surgical decompression, which usually results in a marked improvement of symptoms.

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