C A S E Arachnoiditis ossificans

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Arachnoiditis ossificans is a rare type of chronic arachnoiditis characterised by the presence of calcification or ossification of the spinal arachnoid. There are a few reports of this condition in Japanese and western populations but no case has been reported in a Chinese population before. We describe a 35-year-old woman with typical findings of arachnoiditis ossificans. A brief review of the literature is also presented.

Case report

A 35-year-old woman visited our out-patient clinic complaining of severe low back pain and pain in both lower limbs in June 2007. She had a history of a congenital cervico-thoracic spinal dermoid cyst, managed with multiple operations (laminectomies), the most recent being performed 6 years previously. The spinal lesion caused paraplegia from early childhood but there had been some improvement in her lower limb power after the latest operation, enabling her to walk a few steps with a supporting frame. Nevertheless, she complained of progressive spasticity in her lower limbs over the past 5 years and a progressive decrease in her lower limb power had prevented her from walking for the past three and a half years. She had experienced increasingly severe pain in her lower limbs and lower back region over the past two and a half years. The pain was so severe that she consulted our Emergency Department and was subsequently referred to us for further management. Magnetic resonance imaging (MRI) of the lumbosacral spine was performed. Her spinal MRI showed typical features of arachnoiditis ossificans (Fig 1). There was clumping of the nerve roots in the lumbosacral region consistent with arachnoiditis and calcifications of the thecal sac and the arachnoid around the nerve roots. An abdominal computed tomographic (CT) scan, performed in another institution to investigate her abdominal pain, showed hyperdense calcifications in the lumbosacral region, typical of arachnoiditis ossificans (Fig 2). She declined surgery, so conservative management was offered.

Discussion

Arachnoiditis ossificans is a rare type of chronic arachnoiditis characterised by the presence of calcification in, or ossification of, the spinal arachnoid. It is usually, although not invariably, associated with progressive neurological deficits. The identification of this entity has treatment implications.¹ It usually affects the lumbosacral or the thoracic region; both regions can be affected in the same patient.² No cases of cervical spinal arachnoiditis ossificans have been reported. An association with syringomyelia has been reported.³⁻⁵

Key words Arachnoiditis; Lumbar vertebrae; Ossification, heterotopic; Tomography, X-ray computed

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FIG 1.T1-weighted sagittal magnetic resonance imaging of the lumbar spine showing the intra-thecal arachnoid calcification appearing as a low signal intensity linear structure (white arrow).The retained Myodil from a previous myelogram appears as a small hyperintense nodule (black arrow)



FIG 2.Axial computed tomographic scan at L5 level showing almost circumferential calcification of the dural sac (black arrow) as well as arachnoid calcification around the nerve roots inside the dural sac (white arrow)

Patients commonly present with progressively compressive myelopathy.⁶Thoracic disease symptoms include lower back pain, leg weakness, myelopathy, and signs of spinal cord compression syndrome. With lumbosacral disease, lower back pain, radicular signs with leg pain, sensory disturbances, and bowel and bladder disturbances can occur. Lower limb motor problems, bowel incontinence, urinary frequency or incontinence are common presenting symptoms²; some patients may have mild symptoms.¹

The pathogenesis of arachnoiditis ossificans is not yet understood. It has been proposed that it may be the end stage of adhesive arachnoiditis.⁷ Prior trauma, surgery, subarachnoid haemorrhage, myelography (particularly where oil-based contrast agents have been used), infection, and spinal anaesthesia have all been implicated as causative factors in arachnoiditis ossificans.^{1,26,8}

Our patient has evidence of an intra-thecal Myodil (Glaxo Wellcome, London) injection and spinal surgery, both of which are recognised risk factors for arachnoiditis ossificans. Her progressive deterioration (increasing lower limb spasticity and pain) in recent years suggested the possible development of arachnoiditis ossificans but arachnoiditis ossificans is a rare disease. A literature review published in 1998 found only 46 reported cases in the English literature.² Most case reports describing arachnoiditis ossificans originate from western countries, although a few cases in Japanese patients have been reported. There have been no reports of arachnoiditis ossificans affecting an ethnically Chinese patient in the English literature to date.²

Before the availability of CT, the diagnosis of arachnoiditis ossificans was based on plain X-rays and X-ray tomography. With the introduction of CT, the identification of this disease entity has become much easier.² Although the disease can be identified using MRI, the changes are subtle and may easily be missed.⁹

Due to the overlapping structures and the limited contrast resolution of plain X-rays, it is difficult, if not impossible, to diagnose arachnoiditis ossificans using plain radiography alone.¹⁰ Nevertheless, risk factors for arachnoiditis, such as previous spinal surgery and oily intra-thecal contrast like Myodil, can be readily identified in the X-ray. The evidence of such risk factors and appropriate clinical features should alert radiologists to a possible diagnosis of arachnoiditis ossificans. Myelography alone will show features of arachnoiditis. Nonetheless, the intra-thecal contrast will obscure the arachnoid calcification, rendering it difficult to arrive at the correct diagnosis.¹⁰

The calcifications or ossifications in arachnoiditis ossificans are typically hyperdense on CT scans. They can be identified easily against the

骨化性蛛網膜炎

骨化性蛛網膜炎是一種罕見的慢性蛛網膜炎,特徵為椎管內蛛網膜鈣 化或骨化。日本及西方國家有數個關於此病的報告,但未有類似的華 人病例報導。本報告描述一名35歲女病人的骨化性蛛網膜炎,並簡短 回顧有關的文獻。

hypodense thecal sac and nervous tissue.¹¹ They are usually seen surrounding the spinal cord or thecal sac. In the lumbosacral region, they will also be seen intradurally, adjacent to the nerve roots. The hyperdense calcification or ossification may be mistaken for contrast solution if intra-thecal contrast is being administered as in a myelogram.² Indeed, our patient had a myelogram with CT in another institution and her arachnoid calcification was, unfortunately, missed. A multi-slice CT machine is recommended for investigating patients with arachnoiditis ossificans. The thin slice section and the readily available three-dimensional reconstruction capabilities will greatly facilitate the diagnosis and assessment of the arachnoid calcification.

Computed tomography, with or without intrathecal contrast, is now rarely used to investigate spinal problems because most institutions tend to use MRI for this purpose. Magnetic resonance imaging may show clumping of the nerve roots if the cauda equina region is involved. The nerve root clumping is best demonstrated using axial T2-weighted images. The arachnoid calcification may be difficult to identify using MRI as they have variable signal intensities on MRI. They can appear hypointense or hyperintense in T1-weighted images. In T2-weighted images, the calcifications or ossifications can vary from hypointense to hyperintense.1 Enhancement with gadolinium is also variable. The ossification can appear thin and linear, or globular and mass-like.¹ The use of gradient echo sequences cannot increase the sensitivity of ossification detection due to the presence of chemical shift artifact at the dural-epidural fat interface. Verification of the arachnoid ossification by CT scan is usually required if arachnoiditis ossificans is suspected in the MRI scan.⁵

The prognosis of this disease is variable. Some patients will have progressive compressive myelopathy but some patients remain stable with only mild symptoms, regardless of the degree of ossification. The best treatment strategy for arachnoiditis ossificans has not been established. In patients with severe or deteriorating symptoms, surgery is often performed.⁷ Excision of the dural calcification and microsurgical neurolysis is technically difficult, especially when multiple nerve roots are involved. Even when surgical removal of the intrathecal ossification is possible, the clinical result is generally poor.^{1,7}

Procedures including decompressive laminectomies, anterior fusion, and foraminotomies are preferable and good results have been reported.¹ The surgical option often presents a clinical dilemma. Only 50% of patients managed with an operative intervention showed improvement in the cases reported in the literature.⁵ Surgery itself is a known

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causative factor in arachnoiditis ossificans. In patients with only mild symptoms, conservative management has usually been adopted.^{1,11,12}

Shiraishi et al⁹ has prescribed low-dose aspirin, the aim being to improve local blood circulation, but its effectiveness has not been verified. Steroid therapy, however, is neither helpful nor indicated in this disease.⁹

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