

Rare Causes of Hypertrophic Spinal Pachymeningitis Primarily Identified on Spinal MRI

Dear Editor,

Hypertrophic pachymeningitis is a rare disorder characterized by chronic inflammation and thickening of the dura, which can present with neurological deficits due to compression of the underlying structures. On the basis of the extent of involvement, it can be classified as cranial, craniospinal, or spinal pachymeningitis, of which isolated spinal involvement is the rarest entity. On the basis of the etiology, pachymeningitis can be idiopathic or secondary, associated with various other etiologies like infection, inflammation, granulomatous or neoplastic disorders. The treatment approach for pachymeningitis is not well established, especially in idiopathic cases. Diagnosing the primary cause is crucial for optimal management, which usually requires extensive clinical and laboratory workup. We present two rare cases of hypertrophic spinal pachymeningitis (HSP) associated with Takayasu's arteritis and tuberculosis for which the clue to primary etiology was identified from the initial spinal magnetic resonance imaging (MRI).

The first case is a 52-year-old man who presented with gradually progressive upper thoracic pain and acute onset of difficulty in walking, bowel and bladder incontinence.

Neurological examination showed bilateral ankle clonus and mute bilateral plantar reflex. MRI spine revealed dural thickening suggestive of HSP at T1-T4 levels, causing cord compression and edema [Figure 1a, b, d and e]. Brain screening showed no intracranial dural involvement. The same MRI study revealed incidental findings suggestive of aorto-arteritis [Figure 1c and e]. Pattern of vascular involvement in subsequent contrast-enhanced computed tomography (CECT) [Figure 2] was suggestive of early inflammatory stage Takayasu arteritis. Acute phase reactants were elevated. Serological markers including VDRL and vasculitis profile were negative. In view of progressive myelopathy, canal decompression with dural excision and biopsy was performed, which showed inflammatory changes and vessel wall thickening. The patient was started on low-dose steroids and discharged with normal neurology. Review after 6 months showed complete clinical recovery and no recurrence on imaging [Figure 1f and g].

The second case is a 60-year-old woman who presented with gradually progressive neck pain and bilateral upper limb radiculopathy for one month. MRI revealed dural thickening, suggestive of HSP [Figure 3a and b], extending from C2 to T4 levels with cord compression and edema. The

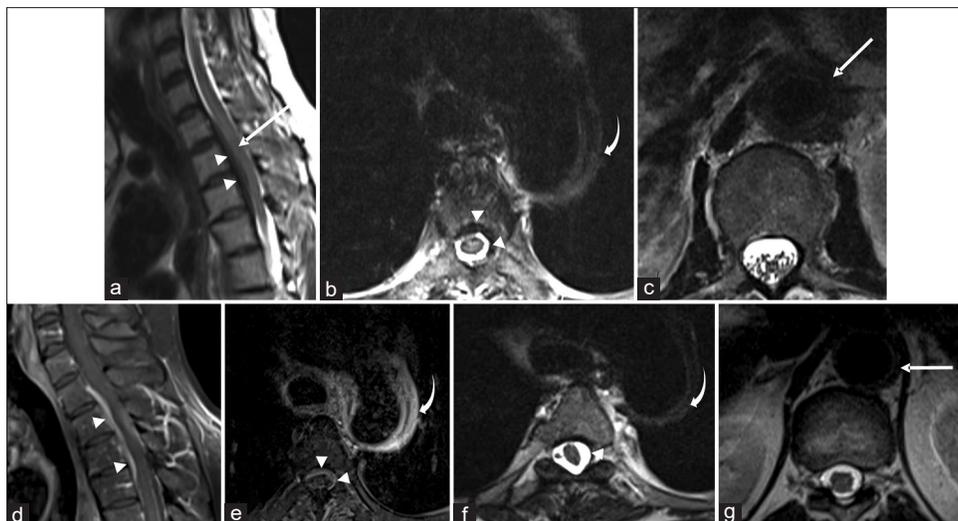


Figure 1: MRI thoracic spine showing HSP and Arteritis. Sagittal and axial T2 weighted (a and b) images of the thoracic spine show diffuse smooth thickening of the anterior and left lateral aspect of the dura (arrowheads in a and b) causing canal stenosis with cord congestion (white arrow in a). The dura is hypointense on T2 and shows homogeneous enhancement on post contrast images (arrowheads in d and e) at T1-T4 levels. Axial T2 weighted (b and c) images at thoracic and upper lumbar levels show concentric wall thickening of the aortic arch (curved arrow in b) and abdominal aorta (white arrow in c). Post-contrast images (e) show enhancement of the aortic arch, suggestive of active aortitis (curved arrow in e). Post-surgical follow-up images (f and g) Axial T2 weighted images of the thoracic spine show complete resolution of dural thickening (arrowhead in g) and significant decrease in aortic wall thickening. (white arrows in f and g)

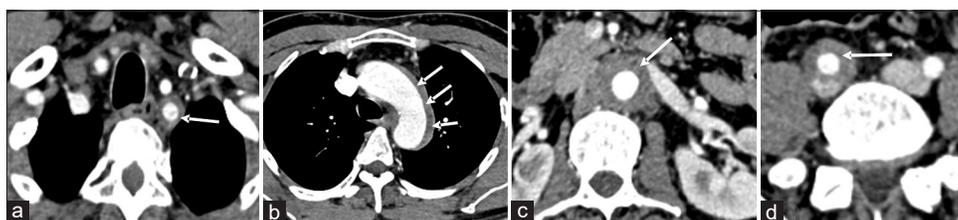


Figure 2: CECT thorax showing features of Takayasu arteritis. Axial, Sagittal and Coronal arterial phase contrast enhanced CT scan of the thorax shows concentric wall thickening of the proximal left subclavian artery (a), arch of aorta (b) and abdominal aorta (c) extending to involve the proximal right common iliac artery (d)

same MRI study also revealed multiple bilateral pulmonary nodules [Figure 3c]. CT-guided biopsy of the pulmonary nodule [Figure 3d] was suggestive of tuberculosis [Figure 3e]. In view of worsening neurological symptoms, canal decompression was performed. Dural biopsy also showed chronic tubercular granulomas.

Spinal pachymeningitis is common in sixth-decade men. Cervical and dorsal levels are more commonly involved. Dural thickening in HSP causes mass effect over adjacent neural structures. Depending upon the level involved and degree of spinal canal stenosis, the patient may present with single or multi-level radiculopathy and myelopathy. Our patients also presented with radiculopathy and myelopathic symptoms.

MRI features of HSP are focal or diffuse dural thickening extending over multiple vertebral levels. Thickened dura is iso or hypointense on T1 and hypointense on T2 weighted images with homogeneous or peripheral rim enhancement on post-contrast images. Since combined craniospinal

involvement is common, imaging of the entire neuraxis has to be performed.^[1] Both of our patients showed classic MRI findings of HSP and brain screening was unremarkable.

There are many reported cases of cranial and spinal pachymeningitis, where the etiology was known before or diagnosed after extensive workup.^[2-4] However, only two cases of Takayasu arteritis with hypertrophic pachymeningitis have been reported in the literature. Kim *et al.*^[5] reported a case of hypertrophic pachymeningitis with spinal involvement, in which the patient was on treatment for chronic peri aortitis before presenting with features of cord compression. Wattamwar *et al.*^[2] reported a case with intracranial dural involvement, which showed classic clinical signs of Takayasu and left subclavian artery stenosis in digital subtraction angiography. However, our patients had no contributory history or clinical signs for the underlying primary cause and the diagnosis were suggested only based on the incidental findings in the initial spine MRI. This was further confirmed by serology and histopathology of the excised dura.

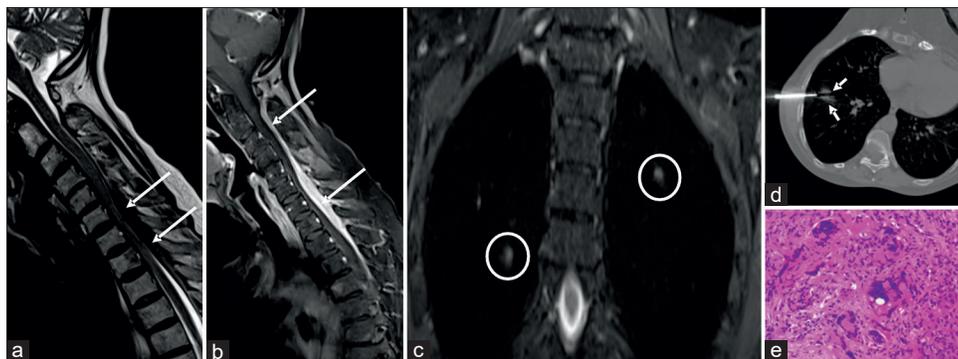


Figure 3: MRI thoracic spine showing HSP with Pulmonary nodules-Histopathologically proven tuberculosis. Sagittal T2 (a) and post contrast (b) images of the cervical and upper thoracic spine show diffuse smooth thickening of the posterior dura (long arrows in a and b) from C2--T4 levels causing canal stenosis with cord congestion. The dura is hypointense on T2 and shows homogeneous post contrast enhancement. STIR coronal images of the thoracic spine showed incidental finding of multiple scattered soft tissue attenuation parenchymal nodules in both lungs (circles in c). CT guided biopsy of the largest nodule in the right lower lobe was performed (short arrows in d). Histopathological examination showed multiple coalescent granulomas with epithelioid histiocytes, Langhans' type multinucleate giant cells, lymphocytes and central areas of incipient necrosis, suggestive of chronic granulomatous inflammation, suggestive of tuberculosis (e)

Tuberculosis is another rare cause of HSP. Most of the cases in the literature had intracranial dural involvement and few of them have reported cervical spinal dural involvement.^[4,6] To our knowledge, there is no reported case of isolated thoracic spinal pachymeningitis due to tuberculosis, where both pachymeningitis and the cause were diagnosed in the initial spinal MRI. Detection of lung nodules in MRI spine helped in early diagnosis of etiology in our patient even before there were systemic symptoms or pulmonary involvement.

Currently, there are no optimal treatment guidelines for hypertrophic pachymeningitis. Idiopathic HSP is often treated with steroids or immunosuppressant. Diagnosing the primary etiology, particularly infection like tuberculosis is important in HSP to avoid post steroid flare-up of underlying infection. Early identification and treatment of underlying diseases like vasculitis, IgG4, Behcets and malignancies might improve treatment outcomes and prevent systemic complications. Our patient with aorto arteritis was given oral steroids and anti-tubercular treatment was started for the patient with tuberculosis.

Early surgical treatment is indicated in HSP to decompress the cord and prevent progressive myelopathy. In view of worsening myelopathic symptoms, both of our patients were surgically decompressed. Post-surgical imaging follow-up is recommended, as high recurrence rates are reported. However, our patients had complete clinical recovery without residual or recurrence on 6 months follow up imaging, probably because the primary cause was also identified and treated simultaneously.

Early surgical and medical management led to complete recovery of our patients, since the primary diagnosis associated with pachymeningitis was suggested based on the opportunistic findings in the initial spinal MRI itself and there was no further delay in management.

Though cord decompression is sufficient for symptomatic relief in HSP, early diagnosis and treatment of the primary cause of HSP might improve treatment outcomes, prevent systemic

complications and avoid recurrence. Often, the primary cause of HSP can be identified in the initial spine MRI with careful evaluation of paraspinal structures in the lumbar spine and lungs, neck spaces in the cervicothoracic spine.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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