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# The Idiopathic Hypertrophic Spinal Pachymeningitis. A case report and review of literature

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## ABSTRACT

Idiopathic hypertrophic spinal pachymeningitis (IHSP) is a rare inflammatory condition characterized by chronic inflammatory hypertrophy of the dura mater. It can involve the entire spine. However, most cases are reported in the cervical and thoracic spine. It can progress from local pain to radiculopathy and eventually develop myelopathy. The aetiology of IHSP is not known. However, it has been suggested to be associated with many diseases. Here we report a case of IHSP in 21-year-old female who presented with paraplegia. The diagnosis was made on MRI Spine and histopathological examination. It was treated with surgical decompression, steroid therapy and patient improved gradually.

## INTRODUCTION

Spinal IHP is a rare disease which causes chronic inflammatory hypertrophy of the duramater. Its etiology is still unknown. It most commonly involves cervical and thoracic spine<sup>1,2,3</sup>. Commonly presents in the age of 6th and 7th decade of life<sup>4</sup>. Symptoms arises due to progressive compression of adjacent structures. We report a rare case of IHSP in a 21 year old women who presented with paraplegia.

## CASE REPORT

A 21 years old female presented with history of progressive weakness of bilateral lower limb since 2½ months, was admitted in department of neurosurgery, SMS hospital jaipur. On examination, modified Ashworth scale of spasticity was grade 2 in bilateral lower limb, Medical Research

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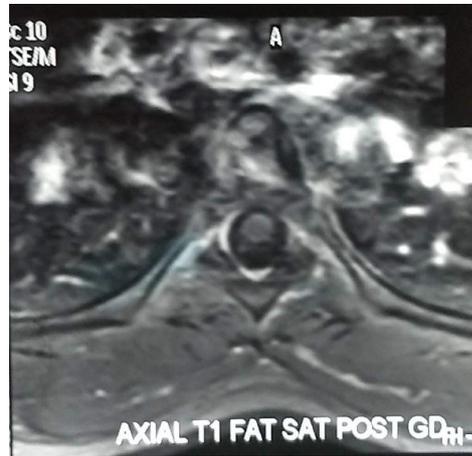


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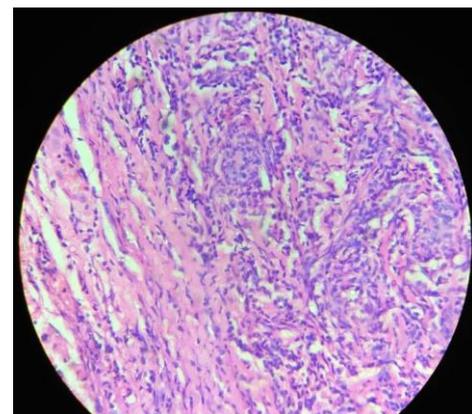
Council power grade was 0/5 in both lower limb, bilateral Knee and ankle reflex was +3. Bilateral Babinski sign was positive and superficial abdominal reflex was absent. 70-80 % sensory loss of all modality below T2 dermatome level was present bilaterally. Other systemic examinations were normal. MR imaging of the thoracic spine demonstrated an intradural extramedullary lesion extending from C7-D4 level that was hyperintense on T1 and hypointense on T2 and nonenhancing on contrast. The lesion was compressing the spinal cord with cord edema. All routine hematological investigations were within normal limit. All the relevant investigations related to the cause of spinal pachymeningitis were negative. After D1–D5 laminectomy, duramater was found to be thickened compressing spinal cord. Posterior excision of duramater was done. A Durafoam was placed posteriorly to expand the thecal sac and specimen was sent for histopathological examinations. Chronic inflammation with lymphoplasmacytic infiltrate and fibrosis was found on histology. Postoperative steroid therapy was given. patient was under follow up and after 8 months, patient developed some sensation in both lower limb and power of lower limb improved from 0/5 to 2/5.



**Figure 1.** MRI Cervico-dorsal Spine sagittal preoperative (A) T1 image- A hyperintense intradural extramedullary lesion present at C7-D4, compressing the spinal cord. (B) T2 image- A hypointense intradural extramedullary lesion present at C7-D4, compressing the spinal cord.



**Figure 2.** MRI Cervico-dorsal Axial image post contrast: (A) Minimal enhancing intradural extramedullary lesion present posterior to spinal cord (B) T2 weighted image- Hypointense lesion present posterior to spinal cord.



**Figure 3.** (A) Intraoperative image showing a grossly thickened dura (B) histopathological examinations showing fibrocollagenous tissue with dense infiltrate of lymphocytes and plasma cells.



**Figure 4.** Postoperative MRI of the cervico-dorsal spine, sagittal view: showing the expanded cord.

## DISCUSSION

This disorder is rare and usually found intracranially and spinal form is extremely rare<sup>5-8</sup>. Friedman D et al found most cases of IHP intracranially<sup>2</sup>. It usually involves the cervical and thoracic dura or occurs as a craniospinal form<sup>(5,9,10)</sup>. The first case of spinal IHSP was reported by Charcot and Joffroy named as "Pachymeningitis hypertrophica cervicalis" in 1869<sup>(11,12)</sup>. Spinal IHSP is a rare cause of nerve root and spinal cord compression. It has been implicated in variety of inflammatory and infectious processes like tuberculosis, sarcoidosis, rheumatoid arthritis, Wegener granulomatosis etc.<sup>(5-8)</sup>. So, other possibilities need to be ruled out as IHSP is a diagnosis of exclusion<sup>(5-7)</sup>. In our case, we could not find any predisposing illnesses such as infectious diseases or autoimmune diseases in spite of thorough investigations. Joffroy and Rosenfeld et al, described three stages; local and radicular pain in first stage. Signs of nerve root compression in second stage and spinal cord compression was described in third stage<sup>(12,13)</sup>. Haobin Chen et al, reported age of the patients ranges from 28 years to 68 years (median age 56 years), and female being more commonly affected (male/female ratio: 6:9). Thoracic spine being most commonly affected

followed by cervical and lumbar spine<sup>14</sup>. Friedman D and Flanders described peripheral enhancement on MRI which was present in all 3 patients as highly suggestive of hypertrophic pachymeningitis, which was also noted in another report<sup>2</sup>. Dumont AS and S.Pai et al, proposed extramedullary mass extending over multiple vertebral levels, strongly hypointense signal on T2-weighted images, and variable peripheral margin enhancement are suggestive of the IHSP<sup>15,16</sup>. In our case, extramedullary lesion was present in C7-D4 level which was hypointense on T2-weighted image and hyperintense on T1-weighted image. spine. Martin N et al reported good result of methotrexate treatment in cranial pachymeningitis<sup>7</sup>. Treatment choices for IHSP include surgical decompression, administration of corticosteroids, radiation therapy, and immunosuppressive agents but relapses are common<sup>(5,17)</sup>. Surgical decompression provides some relief, and early surgical intervention can successfully alleviate neurologic sequelae<sup>1,9,10</sup>. Naffziger and Rosenfeld et al, recommended surgical decompression by laminectomy and excision of the involved dura for its management<sup>(13,15,18)</sup>. Dumont AS and Kitai Ret al reported that biopsy with steroid therapy can reduce the thickness of the dura and can improve neurologic deficit<sup>(15,19)</sup>. Naffziger and Dumont et al suggested laminoplasty instead of extensive laminectomy because it reduces back pain and increases spinal stability<sup>(15,18)</sup>. In our case C7-D4 laminectomy with dural excision was done and post-operative steroid therapy was administered. After 8 months, power of lower limb improved from 0/5 to 2/5.

## CONCLUSION

We reported a rare case of cervico-dorsal compressive myelopathy caused by IHP. Definite diagnosis needs thorough blood investigations, radiological and histopathological findings as IHSP is a diagnosis of exclusion. Surgical decompression along with steroid therapy can be considered as an effective mode of treatment. Early diagnosis and treatment can lead to better neurological outcomes.

## CONFLICT OF INTERESTS

The authors declare no conflict of interests.

## ABBREVIATIONS AND ACRONYMS

IHSP: Idiopathic hypertrophic spinal pachymeningitis; IHP: Idiopathic hypertrophic pachymeningitis; MRI: Magnetic resonance imaging.

**Table 1.** Idiopathic hypertrophic pachymeningitis – Age, location, mode of treatment and its response.

| Author <sup>Ref no.</sup>      | Age | Sex | location                    | Treatment                    | Outcomes   |
|--------------------------------|-----|-----|-----------------------------|------------------------------|--|
| Pai et al <sup>16</sup>        | 68  | F   | Spinal: C6, C7              | Laminectomy, steroid         | spontaneous temporary resolution of symptoms, recurrence after surgery |
| Takahashi et al <sup>20</sup>  | 67  | M   | Spinal C3, C7               | Corticosteroid               | At the 2-year follow-up, the patient could walk independently          |
| Ranasinghe et al <sup>21</sup> | 65  | M   | Spinal T7, T8               | Laminectomy, Corticosteroids | MRI improvement at 57 months   |
| Yasuda et al <sup>22</sup>     | 28  | M   | Spinal: T1, T4, then L1, L3 | Laminectomy, Corticosteroids | Recovered  |
| Lai et al <sup>23</sup>        | 41  | M   | Spinal T2, T4               | Laminectomy, dura excision   | Unavailable  |
| Present study                  | 21  | F   | Spinal C7-D4                | Laminectomy, dura excision   | Partial Recovery   |

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