

# Idiopathic Hypertrophic Spinal Pachymeningitis: A Rare Entity

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## Citation

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## Abstract

We describe a case of idiopathic hypertrophic spinal pachymeningitis in a 62-year-old gentleman presenting with progressive myelopathy of two years. The imaging and corresponding histological findings are discussed and correlated.

## INTRODUCTION

Hypertrophic spinal pachymeningitis (HSP) was first described in 1869 by Charcot and Joffroy (1). Idiopathic hypertrophic pachymeningitis has been described located throughout the central nervous system (CNS). The reported number of cases are still few but the recent increase in numbers of such cases could be attributed to the more frequent usage of gadolinium enhanced MRI (2-4). We report a case of idiopathic hypertrophic spinal pachymeningitis highlighting the histological and radiological findings in light of the current literature.

## CASE

A 62-year-old man presented with progressive weakness of his legs over period of 2 years. He was able to walk with walking aid. The strength in the lower limb was MRC scale of 4/5 bilaterally with brisk reflexes. Examination revealed a sensory level at about T4 dermatome and no sphincter dysfunction. The previous medical history consisted of tonsillar carcinoma 30 years ago, treated with bilateral tonsillectomy and subsequently radiotherapy. He used atenolol 100mg daily for hypertension.

The MRI showed a focal dorsal extradural mass at the level of C7 vertebra. The extradural mass was hyperintense on T2 weighted (W) with a hypointense rim, isointense on T1W.

## Figure 1

Figure 1A: Corresponding sagittal post contrast T1W image with fat suppression reveals dural enhancement of the mass



**Figure 2**

Figure 1B: Sagittal T2W image shows a hyperintense mass with a hypointense rim causing dorsal compression of the spinal cord at C7 level



There was associated cord compression. Post gadolinium fat suppression T1W demonstrated focal dural enhancement. There was loss of marrow with fatty infiltration between C2 to C5 vertebral body due to previous radiotherapy.

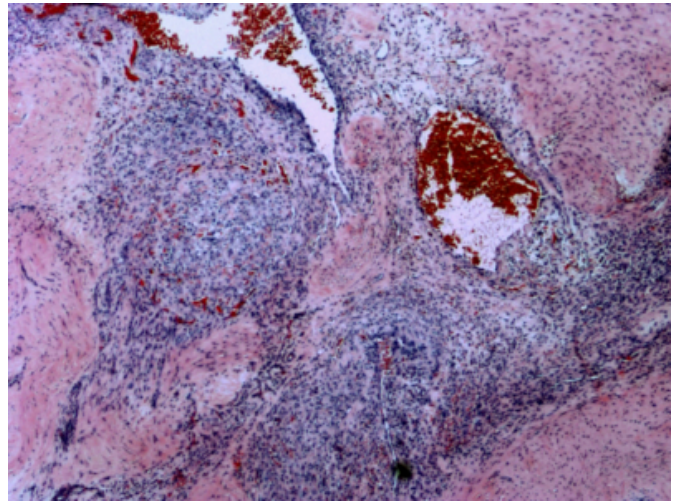
He underwent single cervical laminectomy (C7) with excision of the thickened dura. In-lay duroplasty was done using prosthetic dural material (DURAGEN®) and glued in position with tissue glue (TISSEL®). He was able to walk independently and had improvement in the lower limb sensation 6 months after surgery.

### **PATHOLOGICAL FINDINGS**

The dura was hyalinised, thickened and chronically inflamed with presence of granulation tissue formation.

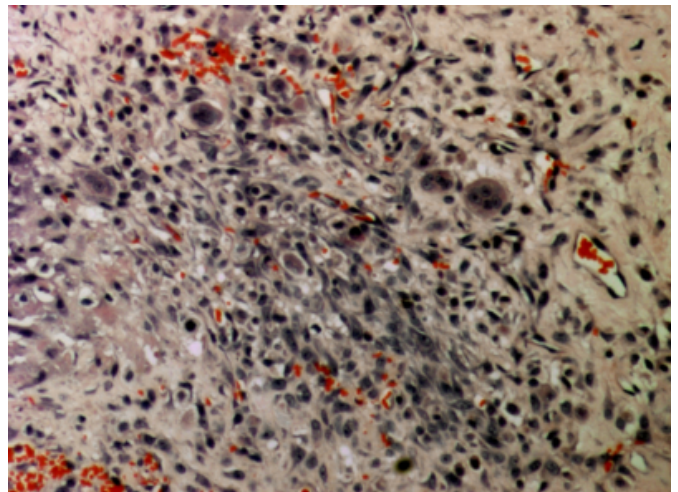
**Figure 3**

FIGURE 2: H&E x 20: Low power view showing lobules of granulation tissue with many dilated and congested blood vessels



**Figure 4**

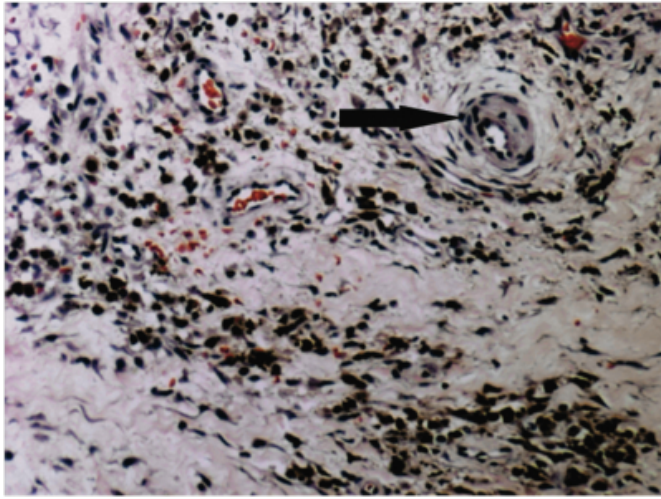
FIGURE 3: H&E x 400: High power view showing the inflamed granulation tissue made up of fibroblastic proliferation, proliferation of small capillaries, scattered chronic inflammatory cells and presence of some foreign-body type multinucleated giant cells



There was no evidence of calcification, malignancy or cyst formation. The hemosiderin-laden macrophages seen in our histopathological examination suggests previous haemorrhage at that site and the chronicity is further strengthened by the presence of small thickened blood vessels.

**Figure 5**

FIGURE 4: H&E x 100: Numerous hemosiderin-laden macrophages admixed with lymphocytes with proliferation of some small capillaries (arrow)



The specimen was also specifically stained for tuberculosis and cultured but investigations were negative. The protein level in the cerebrospinal fluid (CSF) was elevated [2.48 g/L] (normal range 0.15-0.45) but there were no cells or organisms. Extensive preoperative investigation was done, and ESR was slightly elevated to 35mm/hr (normal range 2-10) as was ferritin 1149 µg/L (normal range 22-322). The patient showed no clinical or biochemical evidence of any underlying diseases related to this rare entity.

**DISCUSSION**

Spinal hypertrophic pachymeningitis is caused by chronic inflammation of the dura and the whole spine can be affected (3). The usual location is the cervical or thoracic spine (5). Hypertrophy can be either focal or general, usually only a few levels are affected (one to seven)(2). Involvement of the entire spine has also been described (4). Presentation is not specific and can arise from nerve compression, cord compression or vascular compromise (6). Charcot and Joffroy described 3 stages: the first stage characterized by pain (either local or radicular), the second stage has clinical signs of nerve root compression, and the third stage is characterized by spinal cord compression (7). Early surgical intervention can successfully alleviate neurologic sequelae and laminectomy or laminoplasty followed by durotomy and duroplasty is the recommended surgical treatment for the disease (2,3,6,8). However corticosteroids may achieve symptomatic control and reduction in dural thickness, which can be virtually complete (9,10,11). It has been proposed that hypertrophic spinal pachymeningitis should be considered in the differential diagnosis for patients with

spinal cord compression and radicular pain in more than three spinal levels (5). The clinical course of IHSP may follow 1 of 3 patterns; sustained remission, relapse with corticosteroid resistance or relapse with corticosteroid dependence (12). There are few studies regarding the frequency or cause of recurrence (13).

It has been proposed that a long extramedullary mass of low T2 signal intensity with peripheral enhancement represents a specific MRI finding that is highly suggestive of HSP (6). Our case demonstrates a variable appearance that could occur on T2W in keeping with histopathological findings of previous haemorrhages. The enhancement pattern has been described as either linear or nodular. The linear enhancement pattern appears to show better therapeutic response than the nodular form, possibly related to less fibrosis and more vascularity. These findings are in accordance with the fact that only dura mater and arachnoid mater are affected by miscellaneous chronic inflammation that results in vascularisation and inflammatory cell infiltration. It is difficult on MRI, however, to distinguish these findings from metastatic spinal tumours such as diffuse leptomeningeal carcinomatosis (10).

The histological findings are consistent with the literature stating that most cases of idiopathic pachymeningitis are characterized by a non-necrotizing chronic inflammatory infiltrate of lymphocytes, plasma cells, and occasional histiocytes, giant cells, polymorphonuclear cells, or eosinophils. Granulomas, necrosis and vasculitis are less frequently identified (9,14).

The pathology seen in figure 4 can also be seen after radiation therapy (15), but we believe this to be unrelated as our patients' radiation therapy for tonsillar cancer does not correspond with the location of the pachymeningitis and the lesion is dorsally placed rather than ventral. Furthermore, the radiotherapy changes are only visible in the body of C2 to C5 cervical vertebra. The preoperative work-up is consistent with the existing literature, with elevated ESR, ferritin and CSF protein that is non specific, but likely related to the inflammatory dural process (2).

In an article by Ito et al. analysing HSP, the recurrence rate was 11% over a mean follow-up of 1.3 years. They found factors predisposing recurrence to be at least one positive inflammatory sign before surgery and the duration of the mean follow-up period (3).

In idiopathic HSP the male to female rate is 3:2 and age



distribution range from 15 to 77 years with a mean age of 46 years (13).

The aetiology of hypertrophic pachymeningitis is unknown, but several causative factors have been recognized, among them Wegener's granulomatosis, Mixed Connective Tissue Disease, Systemic Lupus Erythematosus, Sarcoidosis, Multifocal Fibrosclerosis, Orbital Pseudotumor, rheumatoid arthritis, Carcinomatosis, metabolic diseases, trauma, toxins, thrombophlebitis, adjacent to ear or sinus infection, infections such as syphilis, tuberculosis, fungi, HIV, HTLV-1 and meningococcal meningitis and intratecal steroid deposition (2,4,5,9,16). Therefore extensive work up is required to rule out infectious, neoplastic, autoimmune, traumatic, toxic or metabolic cause to diagnose a patient as idiopathic hypertrophic pachymeningitis.

This however can prove more difficult than it sounds as HSP has also been described as initial presentation of vasculitis (12). In the majority of cases the diagnosis is found after surgery (1,8,17-20).

In our case we have not been able to find any causative or related diseases and believe it to be an idiopathic hypertrophic spinal pachymeningitis. One-year follow-up of the patient has still not revealed any signs or symptoms of associated diseases.

## CONCLUSION

Idiopathic hypertrophic spinal pachymeningitis is a rare condition causing symptoms of myelopathy. The variable imaging findings compared to previous publications were due to presence of haemorrhages that was confirmed on histopathological examination. Excision of the lesion and repair of the dura decompressed the spinal cord and brought about good symptomatic relief.

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