CASE REPORT

Hypertrophic pachymeningitis with sarcoidosis: a rare cause of craniocervical compression

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SUMMARY

We describe a case of a 58-year-old woman with a suspected dural tumour. She presented with progressive pyramidal weakness. MRI confirmed compression of the medulla oblongata and spinal cord at the level of C1–3. The localised dural mass lesion homogeneously enhanced on T1 MRI and was considered most likely to be a meningioma. Incidentally, CT scan of the chest revealed peribronchial soft tissue thickening, suggestive of pulmonary sarcoidosis. Owing to the progressive nature of her weakness, she had a posterior occipitocervical decompression with a C1–3 laminectomy and resection of the thickened dura. Histology showed densely collagenous tissue with scanty psammoma bodies and multinucleate giant cells, consistent with hypertrophic pachymeningitis (HP)—a rare, chronic inflammatory condition, characterised by thickening and fibrosis of the dura. This case demonstrates that masses in the craniocervical junction can be varied in pathology and when there is evidence of systemic inflammation, HP should be considered.

BACKGROUND

Hypertrophic pachymeningitis (HP) is a rare, chronic inflammatory condition of the dura. It presents like tumours of the craniocervical junction, which have an insidious onset and cause symptoms of cord and spinal nerve compression when these progress to occupy the subarachnoid space.¹

First described by Charcot and Joffroy,² HP results in dural thickening and opacity, which unites the spinal cord with the dura. Dural inflammation is characterised by the infiltration of plasma cells, macrophages and lymphocytes. The intracranial dura and less commonly, the spinal dura (mainly cervical and thoracic spine) can be affected. There is a slight male to female predominance (3:2) and it commonly affects people in the sixth to seventh decade of life. The condition can be primary (idiopathic) or secondary to infection, inflammatory conditions, neoplasia, trauma and toxins.^{2 3}

CASE PRESENTATION

We present a case of cervical HP in a 58-year-old woman, previously fit and well, who presented with slow onset pyramidal muscle weakness, worse in the upper limbs. There was no lymphadenopathy and no temporal artery tenderness.

INVESTIGATIONS

The patient's full blood count, renal function and liver function tests were normal, but the inflammatory markers were raised (C reactive protein (CRP)

32 mg/L, erythrocyte sedimentation rate, (ESR) 70 mm/h). No infective cause was found; hepatitis and Lyme disease serology were negative and there was no growth from blood or urine cultures. An autoimmune screen was negative and there was no evidence of neoplasia. Her serum ACE level was 19 U/L (8–65 U/L).

The chest X-ray was normal, but there was evidence of peribronchial soft tissue thickening with atelectasis of the left lower lobe on CT thorax. This was suggestive of sarcoid, but the patient declined bronchoscopy for further confirmation. Liver ultrasound was unremarkable.

Contrast enhance MRI of the head and cervical spine demonstrated thickening of the dura extending from the foramen magnum to the level of the C2–3 disc; this was more marked dorsally and lateral to the cord (right more than left). There was no cord compression or signal change in the cord. Initially, an *en plaque* meningioma was considered to be the most likely diagnosis (figure 1).

TREATMENT

The patient underwent a posterior occipitocervical decompression with C1–3 laminectomy and removal of the posterior edge of the foramen magnum. The underlying ventral dura was thickened to the level of C3 and it was incised to reveal a normal arachnoid layer and underlying cervical spinal cord. The dura was tough and it was necessary to decompress the cervical spinal cord by cutting away the meningeal thickening with a scalpel. A dural patch was inserted and the wound was closed with absorbable sutures.

OUTCOME AND FOLLOW-UP

Histology showed benign densely collagenous tissue with scanty psammoma bodies and numerous multinucleate giant cells. These findings were suggestive of chronic pachymeningitis (figure 2).

Postoperative course

The patient's myelopathic symptoms, fine finger function and balance all improved. She was not started on corticosteroids postoperatively.

DISCUSSION

Charcot and Joffroy² described three stages of HP. First, the patient suffers from local and progressive radicular symptoms as a result of nerve root compression. Second, muscle weakness and atrophy follow. Third, the patient experiences symptoms related to spinal cord compression as a result of dural thickening and impending pressure. This is

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Figure 1 On the left is a sagittal post gadolinium T1-weighted preoperative MRI of the cervical spine demonstrating dural thickening and enhancement from the foramen magnum to the level of the C2–3 disc. On the right is a sagittal post gadolinium T1-weighted postoperative MRI.

characterised by weakness, paraplegia, urinary and faecal incontinence and respiratory distress.^{2 4}

Our patient's histology is consistent with HP, characterised by multinucleate giant cells, psammoma bodies and dense collagenous fibrosis. The key pathological feature of HP is dural thickening caused by non-necrotising inflammation and followed by dense collagenous fibrosis, which is considered a terminal event in the disease process. For reasons which remain unknown, there is infiltration by inflammatory cells; lymphocytes, plasma cells and less commonly, histiocytes, giant cells and eosinophils. Dural biopsy is, therefore, the key to establish the diagnosis.^{5 6}

Rossi *et al*,⁶ found elevated markers for CD3+ and CD4+ T cells in 4 patients with HP. They suggest this infiltration of lymphocytes may result from a cell-mediated immune process triggered by antigens or auto-antigens. Riku *et al*,⁵ found IgG4 positive plasma cells on histology when reviewing autopsy samples taken from patients with HP. They suggest HP may be an IgG4-related systemic disease.⁶

We investigated secondary causes of HP based on causes documented in current literature. These include infection, inflammatory conditions, neoplasia, trauma, and toxins. On admission, our patient's serum ACE, cytoplasmic antineutrophil cytoplasmic antibodies and perinuclear antineutrophil cytoplasmic antibodies (p-ANCA), rheumatic factor and antineutrophil antibodies were all within normal limits.^{2 3}

Figure 2 Photomicrograph showing multinucleate giant cells and psammoma bodies. H&E stain. Medium power magnification.

Similar findings were reported by Kupersmith *et al*⁸ in a review of 12 patients with idiopathic HP. However, Takuma *et al*¹⁰ reported p-ANCA positive HP. Granulomatous changes have also been reported in cases of HP. As both p-ANCA antibodies and granulomatous changes are detected in vasculitides, there is a suggestion that microvasculitis may be involved in HP. Further support for the involvement of microvasculitis in HP comes from studies that have shown raised inflammatory markers. Our patient's ESR and CRP were raised. Kim *et al*¹⁷ reported similar findings in their two patients with HP, Parney *et al*⁹ in their patient and Kupersmith *et al*⁸ reported that 5 of their 12 patients had raised ESR.^{7–11}

Our patient's MRI, with gadolinium of the craniocervical junction, showed an enhancing thickening of the dura extending from the foramen magnum to the level of the C2–3 intervertebral disc. Pai *et al*¹² report similar MRI findings in two patients with HP; the MRI showed peripheral enhancement of an extramedullary mass which encompassed and compressed the spinal cord. In a review of the literature, they found that 12 of 14 published cases of HP reported similar MRI findings. They proposed that this was a specific MRI finding that is highly suggestive of HP. The enhancement of an extramedullary mass is likely to represent the chronic inflammation and fibrosis of the dura mater. We note that the dorsal dura was more thickened compared with the ventral dura, findings also reported by Ranasinghe *et al*¹³ who described thickness in the dorsal dura varying from 5 to 15 mm in a study of three patients with HP.^{12 13}

The availability of MRI has led to an increase in the detection of HP. HP is generally reported as dural thickening over multiple spinal levels, enhancing uniformly and appearing either isointense or hypointense on T1-weighted and T2-weighted images.¹⁴

Surgical decompression is used to diagnose HP and provide symptomatic relief. However, recurrence of HP is common. Ito *et al*¹⁵ reviewed 96 spinal HP case reports and found 11 patients had recurrence of HP. They suggested recurrence of HP is significantly greater in patients who had active dural inflammation preoperatively and residual arachnoiditis.¹⁵

Many authors report use of low dose lifelong corticosteroids, radiotherapy and immunosuppressants, such as azathioprine, to prevent recurrence postoperatively. Kupersmith *et al*⁸ reported that 10 of their 12 patients had improved neurological function after being treated with corticosteroids. They also report relapse when tapering the dosage of steroids. Bosman *et al*¹⁶ identified 60 treated cases of idiopathic HP when they carried out a search for English-language literature using the PubMed database. They reported that 93% of the 60 patients were treated with corticosteroids. They also reported that patient's receiving corticosteroid as monotherapy were more likely to relapse. They found patients receiving add-on therapy, which include azathioprine, cyclophosphamide or methotrexate,

Learning points

- ▶ Hypertrophic pachymeningitis (HP) can only be confirmed by histology.
- ▶ The cause of the dural inflammation in HP is still unclear.
- ▶ Patients diagnosed with HP should be screened for secondary causes such as infection, inflammatory conditions, neoplasia, trauma and toxins.

had a sustained remission allowing tapering of the corticosteroid dose.^{7 8 16 17} We elected not to administer corticosteroids post-operatively in view of the potential long-term side effects and evidence of relapse due to tapering of steroids.

Competing interests None declared.

Patient consent Obtained.

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