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Case Report

Craniocervical hypertrophic pachymeningitis

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ABSTRACT

Background: Hypertrophic pachymeningitis (HP) is a rare neurological disorder characterized by dural thickening. Here, we discuss the diagnosis and surgical management of a 38-year-old whose myelopathy was attributed to dorsally compressive HP extending from the lower cerebellar fossa to C3.

Case Description: A 38-year-old male with Sjögren's syndrome presented with cervical pain, upper limb paresis, dysphagia, and left-sided tongue/palate paralysis. The cervical magnetic resonance (MR) showed a dorsally compressive lower cerebellar fossa to C3 lesion. When the biopsy revealed HP, and once conservative treatment failed, the patient successfully underwent a posterior surgical decompression, lesion debulking, and craniocervical fusion.

Conclusion: Cervical HPs should be diagnosed early on MR, and those with significant myelopathy, aggressively surgically treated.

Keywords: Case report, Craniocervical junction, Dural thickening, Hypertrophic pachymeningitis, Meningitis

INTRODUCTION

Hypertrophic pachymeningitis (HP) rarely involves thee spine. Chronic HP, due to progressive thickening/fibrosis of the dura mater, can contribute to brain/spinal cord compression.[8,10] Magnetic resonance (MR) findings are not sufficient to establish the diagnosis of HP. Rather, biopsy confirmation of HP is essential to confirm HP and differentiate it from syphilis, tuberculosis, fungal infections, autoimmune diseases, and Wegener's granulomatosis.[5] Here, we reviewed the diagnostic and therapeutic challenges for a patient presenting with a craniocervical junction HP lesion.

CASE DESCRIPTION

A 38-year-old male patient presented with 4 months of increased difficulty speaking due to left-sided tongue/palate paralysis a progressive quadriparesis. The cervical MR imaging

(MRI) showed an expansive/dorsally compressive dural broad-based posterior cranio-cervical junction lesion extending from the lower cerebellar fossa to the C3/C4 level. The dorsal dural lesion was 9 mm thick in its largest anteroposterior diameter and displaced the spinal cord anteriorly. Further, the cranial MRI revealed regular, concentric, and homogeneous thickening of the pachymeninges that extended from the anterior sella turcica along the entire ventral aspect of the brainstem [Figure 1].

Biopsy and immunohistochemical analysis

The biopsy and immunohistochemical analyses of the lesion revealed chronic lymphoplasmacytic meningitis (i.e., focally granulomatous, without acid-fast bacilli or fungi on special stains). Cellular markers confirmed the presence of B and T lymphocytes and macrophages, suggesting a chronic inflammatory process. However, there was no evidence of a syphilitic infection, and no Langerhans cells were present [Figure 2].

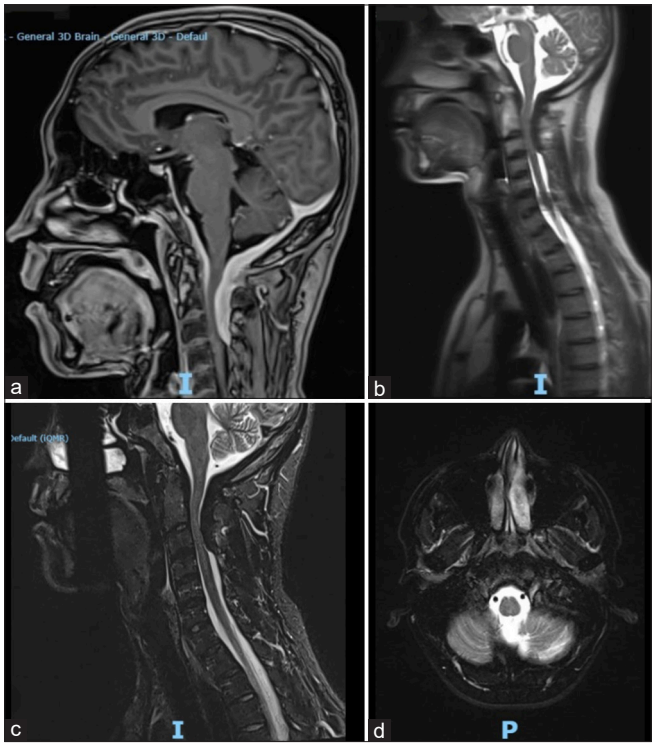

Figure 1: Preoperative magnetic resonance (a) sagittal T1-weighted spoiled gradient echo magnetic resonance imaging (MRI) for high-resolution anatomical imaging, (b) localizing scan in three planes (axial, coronal, and sagittal), (c) short-tau inversion recovery MRI of T1 spine, highlighting soft tissues/edema, and (d) axial section of T2 level.

Craniocervical junction HP surgery

The patient failed conservative treatment and required a posterior fossa/C1–C3 craniectomy/laminectomy; it was performed under intraoperative neurophysiological monitoring using total intravenous anesthesia. At surgery, the HP lesion appeared as a thickening of the dura mater [Figure 3]. The patient underwent partial resection of the HP lesion to relieve suboccipital, cervicomedullary, and upper spinal cord compression as confirmed on postoperative brain/cervical MR studies [Figure 4].

DISCUSSION

Nodular HP is a rare chronic inflammatory process that involves cranial and/or spinal dural fibrosis.[1,3] It may present as idiopathic or with secondary causes variably attributed to rheumatoid arthritis, immunoglobulin G4-related disease, sarcoidosis, Sjögren's syndrome, neoplasms, and infectious etiologies. Symptoms may include cranial (i.e., headaches and increased intracranial pressure) and/or spinal (i.e., radiculopathy/myelopathy) complaints. Here, our patient presented with left-sided paralysis of cranial nerves X and XII and quadriparesis. The MR with/without contrast documented HP cranial/spinal low T2 signal lesions compressing the brain/cord.[6,7] The biopsy, a gold standard for diagnosing HP lesions, documented HP that was resected utilizing a suboccipital craniectomy/laminectomy C1–C3 with fusion.[4] The literature confirms that the best therapeutic approaches typically include direct surgical resection of the HP mass.[1,2,9] Here, the 5-month post-operative MRI documented successful HP lesion partial resection without regrowth [Table 1].

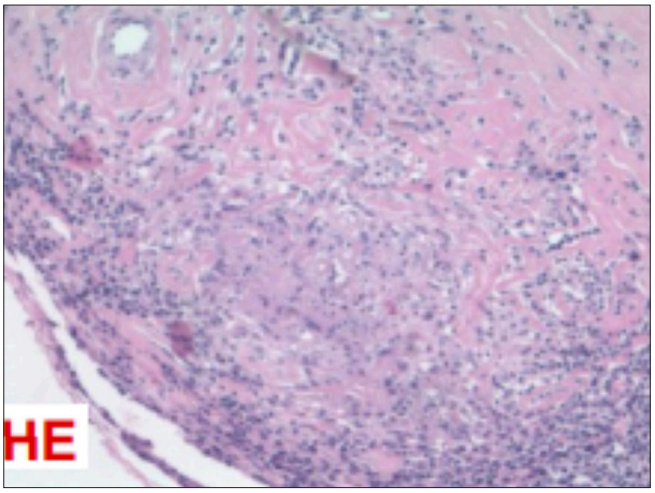

Figure 2: Immunohistochemistry revealing chronic inflammation with B/T lymphocytes and macrophages; no syphilitic infection or Langerhans cells detected.

Table 1: Characteristics of the included references.

Author	Journal	Year	Data/Design	Findings	Conclusion
Bang *et al.*[1]	Korean J Spine	2015	Case Report	HP CCJ/SCS	Surgery Required
Choi *et al.*[2]	Korean J Spine	2008	Case Series	5 Surgery 3 Recurrences	Surgery Effective Risk Recurrence
D'Andrea *et al.*[3]	Neurosurg Rev	2004	Case Report Review	IIHP	Steroids Surgery
Hamada *et al.*[4]	Neurosurgery	2000	Case Report	Cranial HP Plus Dural AV Fistula	Vascular Studies Needed
Jee *et al.*[5]	J Korean Neurosurg Soc	2014	Case Report	HP-Osteolytic Lesion; Biopsy chronic inflammation	Imaging Needed for Diagnosis
Lee *et al.*[6]	AJNR Am J Neuroradiol	2003	Case Report	Progressive Meningeal Thickening 7-yr F/O	Long-term F/O Needed
Pai *et al.*[7]	AJNR Am J Neuroradiol	2007	Two Cases	Two Cases IIHP Surgery	Surgery Effective
Qin *et al.*[8]	Eur Spine J	2015	Case Report Review	IIHP	Broad Differential Diagnosis, Need Biopsy
Ranasinghe *et al.*[9]	J Neurosurg Spine	2011	Case Series	IIHP Neurological Deficits	IIIHP Multimodal Treatment Long-term F/O
Zhu *et al.*[10]	Eur Spine J	2015	Case Report	Craniocervical IIHP	Diagnosis MR-Biopsy Needed

CCJ: Craniocervical junction, SCS: Spinal cord compression, IIHP: Idiopathic intracranial hypertrophic pachymeningitis, HP: Hypertrophic pachymeningitis, yr: Year, F/O: Follow-up, MR: Magnetic resonance

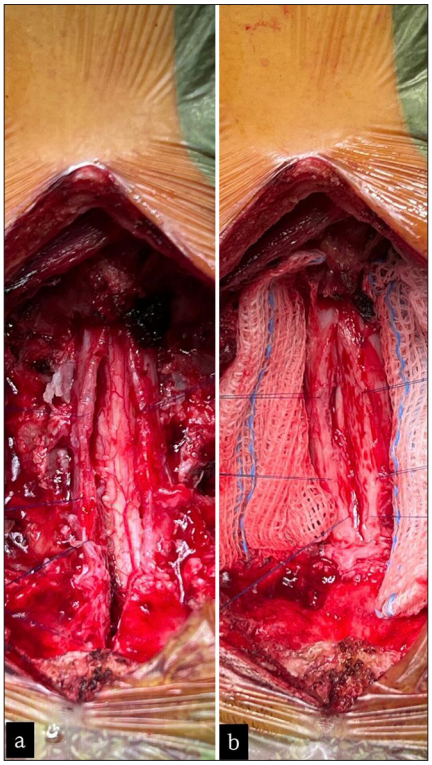

Figure 3: (a) Postoperative photograph and (b) preoperative photograph showing thickened dura.

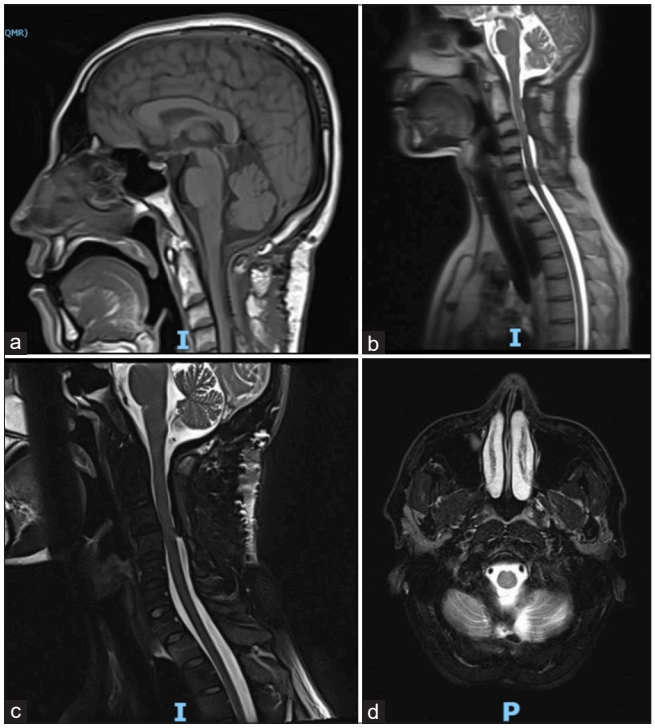

Figure 4: Postoperative cervical Magnetic resonance. (a) Sagittal T1-weighted Magnetic resonance imaging (MRI) brain, (b) localizer scan in three planes (axial, coronal, and sagittal), (c) sagittal section of T2 level, and (d) axial section of T2 level.

CONCLUSION

HP rarely causes cranial/spinal dural thickening and brain/cord compression. Establishing the correct diagnosis with MR, obtaining biopsy confirmation of the HP lesion, and following with partial/gross total HP tumor resection is critical to optimize neurological improvement/recovery.

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