Commentary

Hypertrophic pachymeningitis refers to a clinical disorder characterized by diffuse thickening of the dura mater with or without associated inflammatory changes seen on histopathology.[1] The case report titled "Idiopathic hypertrophic pachymeningitis presenting with a superficial soft tissue" by Keshavaraj *et al*.[2] describe a 40-year-old woman who presented with chronic headaches and a facial mass. Magnetic resonance imaging (MRI) of the brain revealed meningeal thickening and meningeal biopsy revealed fibroconnective dural tissue with chronic inflammatory infiltrate. Further workup for rheumatologic, infectious, and neoplastic disorders was unrevealing. These findings are consistent with idiopathic hypertrophic pachymeningitis (IHP) presenting with an unusual focal facial mass.

IHP is a rare disorder affecting predominantly males with prevalence in the 6th decade of life.[3] Upon review of the literature, there have been a number of case reports found describing the diverse neurological manifestations and anatomical localizations of this peculiar disorder. These have included idiopathic tumefactive hypertrophic pachymeningitis presenting as a large frontal lobe mass,[3] IHP of the thoracic dura mater presenting with a sensory level at T8,[4] IHP affecting the cranial nerves presenting with transient hemianopsia,[5] and IHP presenting with

bilateral papilledema, headaches, and fevers with dural sinus thrombosis and pachymeningeal thickening seen on MRI and MRV of the brain.[6]

Of the presenting symptoms of cranial IHP, headaches most commonly occur in 92% of patients followed by vision loss in 58% of patients.[1] Other neurological manifestations may include seizures, cerebellar ataxia, hydrocephalus, and optic neuropathy.

IHP has been seen in association with other syndromes. Cano *et al*. presented a case report of cranial IHP with Sweet's syndrome.[7] Other conditions described in the literature include Tolosa–Hunt syndrome, cranial polyneuritis, thyroiditis, multifocal fibrosclerosis, and diabetes insipidus.[3] A case of IHP has been found in association with myocarditis responsive to steroids suggesting a common autoimmune etiology.[8]

Initial treatment of IHP includes corticosteroids which have shown to effectively arrest the clinical progression of the condition. Relapse rates may be observed in up to 66% of the cases. Cases that are refractory to steroids may require an immunosuppressive agent such as azathioprine which may be used with or without decompression surgery.[9]

This case, presenting with a soft and superficial facial mass histopathologically consistent with chronic inflammation, illustrates the diverse presentation of this uncommon disorder. IHP may constitute a diagnostic dilemma due to its rarity, and variable presentation and localization. Meningeal biopsy is useful in establishing the diagnosis in order to start prompt treatment.

In conclusion, IHP manifestations may include ataxia, headaches, vision loss, cranial neuropathy, tumefactive mass, dural sinus thrombosis, sensory level, hydrocephalus, and seizures among other neurological findings. The authors of the present case describe a focal facial inflammatory mass as the initial presentation of IHP. This condition should also be considered when clinicians encounter patients with a history of headaches, an inflammatory mass, and diffuse dura thickening on brain imaging.

Max R Lowden

Department of Neurology, The Pennsylvania State University Milton S. Hershey Medical Center, College of Medicine, USA

Address for correspondence:
Dr. Max Lowden,
Department of Neurology, Penn State Hershey Medical Center,
30 Hope Drive, PO Box 859, Hershey, PA 17033, USA.
E-mail: mlowden@hmc.psu.edu

References

1. Kupersmith MJ, Martin V, Heller G, Shah A, Mitnick HS. Idiopathic hypertrophic pachymeningitis. Neurology 2004;62:686-94.
2. Keshavaraj A, Gamage R, Jayaweera GM, Gooneratne IK. Idiopathic hypertrophic pachymeningitis presenting with a superficial soft tissue mass. J Neurosci Rural Pract 2012;3:193-5
3. Kazem IA, Robinette NL, Roosen N, Schaldenbrand MF, Kim JK. Idiotumefactive hypertrophic pachymeningitis. Radiographics 2005;25: 1075-80.
4. Lowden MR, Gill D. Teaching NeuroImage: Idiopathic hypertrophic spinal pachymeningitis. Neurology 2009;72: E27.
5. Bosman T, Simonin C, Launay D, Caron S, Destée A, Defebvre L. Idiopathic hypertrophic cranial pachymeningitis treated by oral methotrexate: A case report and review of literature. Rheumatol Int 2008;28:713-8.
6. Goyal M, Malik A, Mishra NK, Gaikwad SB. Idiopathic hypertrophic pachymeningitis: Spectrum of the disease. Neuroradiology 1997;39:619-23.
7. Cano A, Ribes R, de la Riva A, Rubio FL, Sánchez C, Sancho JL. Idiopathic hypertrophic cranial pachymeningitis associated with Sweet's Syndrome. Eur J Radiol 2002;44:139-42.
8. Tanaka M, Suda M, Ishikawa Y, Fujitake J, Fujii H, Tatsuoka Y. Idiopathichypertrophic cranial pachymeningitis associated with hydrocephalus and myocarditis: Remarkable steriod-induced remission of hypertrophic dura mater. Neurology 1996;46:554-6.
9. Lee YC, Chueng YC, Hsu SW, Lui CC. Idiopathic hypertrophic cranial pachymeningitis: Case report with 7 years of imaging follow-up. AJNR Am J Neuroradiol 2003;24:119-23.

Access this article online	
Quick Response Code:	Website: www.ruralneuropractice.com