Case Report

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Night View of Eiffel Tower'- Radiological Sign of Chronic Idiopathic Hypertrophic Pachymeningitis: A Case Report

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ABSTRACT

Chronic idiopathic hypertrophic pachymeningitis (IHP) is a rare inflammatory entity with unknown etiopathogenesis, resulting in dural thickening and fibrosis. This mimics the illuminated night view of the Eiffel tower on contrast-enhanced MRI. The authors report a characteristic MRI sign of chronic IHP on contrast-enhanced MRI. The authors present a case of a 30-year-old male presenting with a headache for the past 3 years with recent aggravation of symptoms. Laboratory and imaging results were reviewed. Follow-up imaging with ongoing treatment was also reviewed. The etiology could not be determined even after extensive investigations. Contrast-enhanced MRI revealed diffuse enhancing thickening of the pachymeninges along the posterior cerebral hemispheres, falx cerebri, and tentorium cerebelli, giving the typical 'Night view of the Eiffel tower' appearance on coronal images. There was cerebral venous sinus thrombosis secondary to IHP. The patient was treated with corticosteroids and anticoagulants. The follow-up scan revealed reduced thickness and enhancement of the meninges.

Introduction

Idiopathic Hypertrophic pachymeningitis (IHP) is a rare inflammatory entity with unknown etiopathogenesis, resulting in dural thickening and fibrosis [1]. It is a diagnosis of exclusion of secondary causes such as infection, malignancy, and systematic autoimmune diseases [2]. There are various presentations like chronic headache, ataxia, and cranial neuropathies [3]. The association of dural venous sinus thrombosis/occlusion is rare. The authors report a case of chronic IHP and dural sinus thrombosis giving a peculiar MRI sign 'Eiffel-by-Night' or 'Night view of Eiffel tower' appearance on contrast study. Follow-up imaging was also reviewed with ongoing treatment on corticosteroids and anticoagulants.

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Case History

A 30-year-old man had several episodes of headache for the past three years. His symptoms aggravated over the past few months with minimal nuchal rigidity and low-grade fever. There were no other neurological symptoms. The patient had no history of diabetes, hypertension, or any other long-term illness. The patient underwent a clinical examination where he revealed no neurological deficits. There was no evidence of lymphadenopathy, no organ enlargement, or other systemic features. A panel of cerebrospinal fluid (CSF) and blood analysis was conducted to rule out infectious, immune, or neoplastic etiology of the patient's symptoms. Laboratory tests showed an elevated erythrocyte sedimentation rate (ESR) (56 mm/h) and C-reactive protein (CRP) (80 mg/L). Serum and CSF tests were negative for antinuclear antibody series, Anti-neutrophil cytoplasmic antibodies, rheumatoid factors (RF), complement (C3/C4), and immunoglobin G (IgG). Magnetic resonance imaging (MRI) with contrast revealed diffuse enhancing thickening of pachymeninges along the posterior cerebral hemispheres, falx cerebri, and tentorium cerebelli (Figure 1), giving the typical 'Eiffel-by-Night' appearance on coronal images (Figure 2). There was cerebral venous sinus thrombosis/occlusion (CSVT/CSVO) confirmed on MR venography of the superior sagittal sinus, straight sinus, and bilateral transverse and sigmoid sinuses (Figure 3). There was no involvement of brain parenchyma or cranial nerves. The patient was treated with corticosteroids (methylprednisolone 1 mg/kg body weight/day with gradual tapering) and anticoagulants (injection Enoxaparin 1 mg/kg subcutaneous route twice daily with warfarin overlap, discharged on warfarin with weekly INR monitoring). He improved symptomatically; his headache resolved. He was rescanned after three months. Repeat MRI suggested reduced dural (pachymeningeal) thickness and enhancement (Figure 4). CSVT persisted in the repeat scan with no evidence of recanalization.

Discussion

Chronic IHP is a rare dural inflammatory process with unknown etiology [1]. It was first described by Charcot and Joffroy in 1869 [4]. There are various causes of pachymeningeal enhancement such as autoimmune, neoplastic, and infectious causes. 'Idiopathic' is a term when all the above causes are excluded [1-3]. Clinical presentation is highly variable ranging from headache, cranial neuropathies, ataxia, seizures,

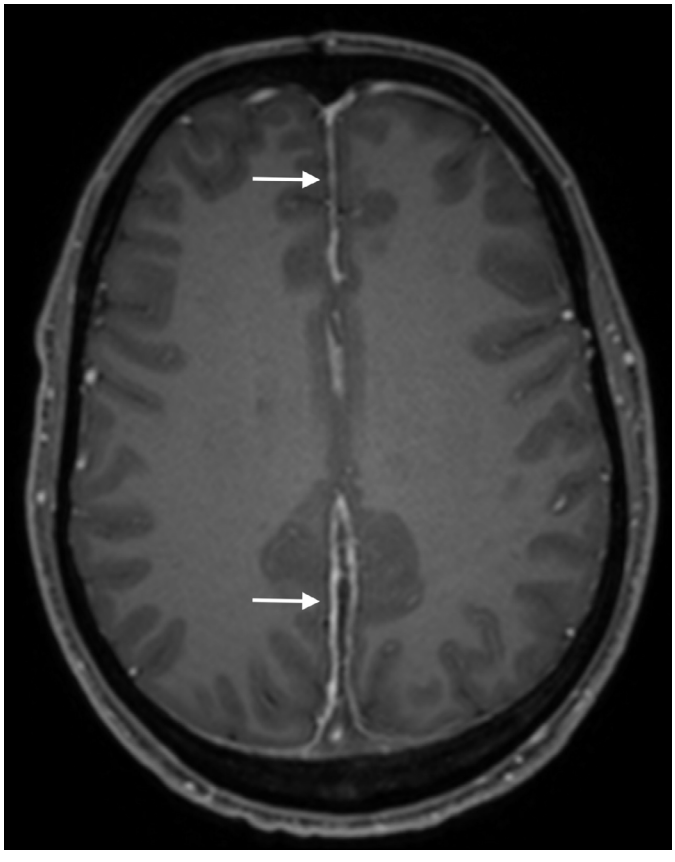

Fig. 1. T1 weighted axial post-contrast image showing pachymeningeal enhancement along falx.

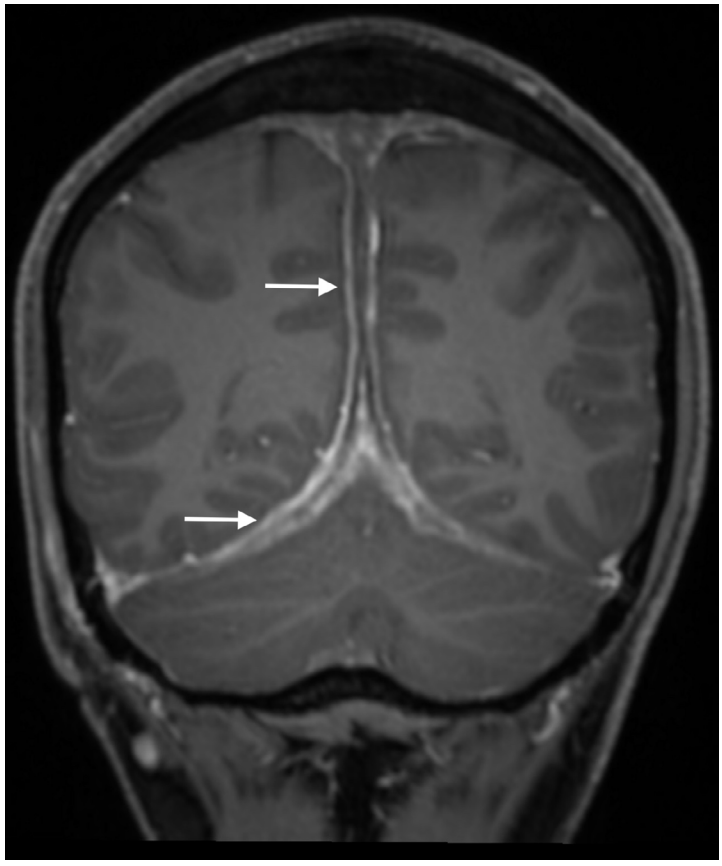

Fig. 2. T1 weighted coronal post-contrast image showing pachymeningeal enhancement along falx and tentorium -mimicking 'Eiffel-by-Night' appearance.

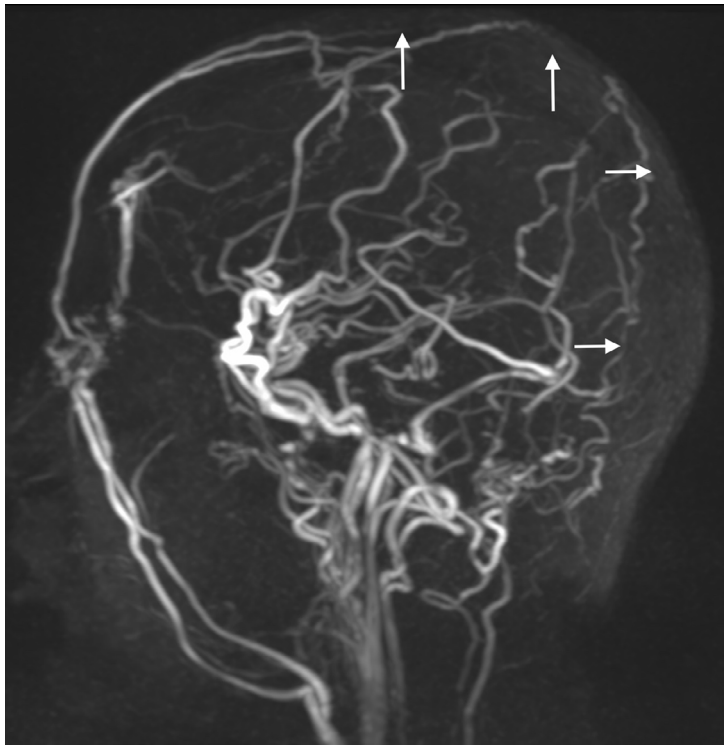

Fig. 3. MRV sagittal reconstructed images suggesting filling defect in dural venous sinuses

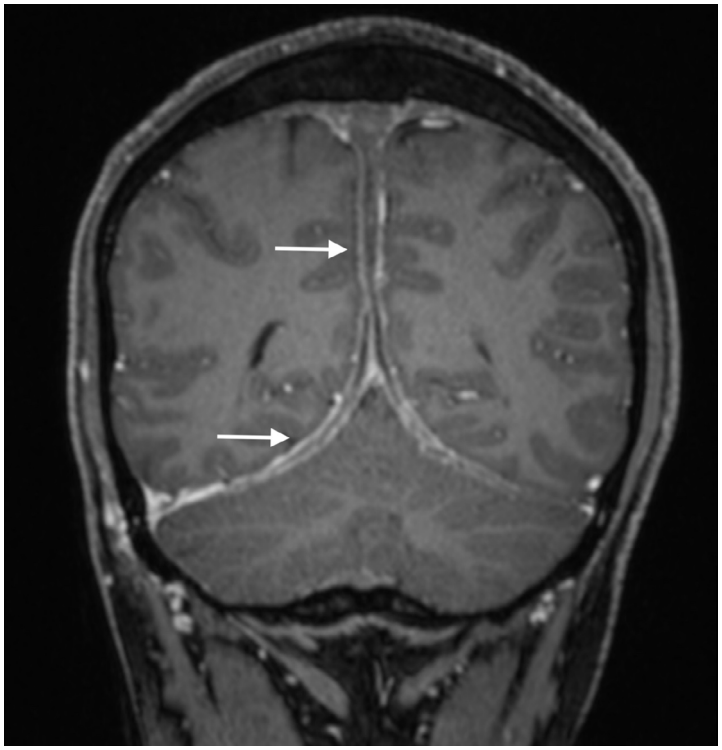

Fig. 4. T1 weighted coronal post-contrast image showing reduced pachymeningeal enhancement along falx and tentorium

psychiatric disorders, features of raised intracranial pressure due to mechanical compressive changes on intracranial structures due to meningeal thickening. The most common symptom being headache likely due to dural inflammation [3]. The incidence of dural venous sinus thrombosis/occlusion with IHP is even rarer [5,6].

The contrast-enhanced MRI reveals diffuse pachymeningeal thickening and enhancement. On coronal scan, the involvement of the posterior aspect of falx cerebri and tentorium cerebelli gives a typical 'Eiffel-by-Night' or Night view of 'Eiffel tower' sign [7,8]. Progressive fibrosis of the dural layer leads to encasement of dural sinuses leading to CSVT/CSVO. The thrombosis of the sagittal sinus (62%) and bilateral transverse sinus (86%) is the most common occurrence with IHP according to the International Study on Cerebral Vein and Dural Sinus Thrombosis (ISCVDST). There could be various reasons for the above finding. The involvement of falx and tentorium is more in IHP, and the affected sinus lies next to them. Another explanation could be the prethrombotic state of IHP which probably affects the sinuses.

Clinically, patients with IHP and CVST/CVSO have a higher incidence of severe headache, intracranial hypertension, psychiatric abnormality, meningeal signs, and seizures [5,6,9]. The authors' patient also had severe headache as well as nuchal rigidity (meningeal sign). According to the ISCVDST study, about half of the patients with sinus thrombosis develop parenchymal lesions and neurological signs [9]. This was not found in the authors' case. For the management of IHP, multiple published reports have shown the effectiveness of the use of glucocorticoids as the initial therapy [10]. The use of immunosuppressants including azathioprine [3], oral [2] or subcutaneous methotrexate [11] cyclophosphamide, and recently, intraventricular cytarabine, have also been tried [12].

The use of anticoagulants for IHP with sinovenous occlusive disease is new. Although, it is less effective for managing occlusion due to sclerosis and is mainly effective if there is thrombosis. The European Federation of Neurological Sciences guidelines published in 2006 detail the use of anticoagulants in patients with acute cerebral venous sinus thrombosis (CVST) [13]. The authors' patient was treated with glucocorticoids and anticoagulants and showed clinical improvement on treatment. The authors' patient highlights the classical 'Eiffel-by-Night' appearance on coronal contrast-enhanced MRI images. This radiological sign helps clinch the diagnosis of hypertrophic pachymeningitis.

Additionally, it is essential to look for complications of IHP like CVST/CVSO in patients presenting with chronic headache and meningeal signs, as seen in the authors' case.

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this article.

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Conflict of Interests

The authors have no conflict of interest to declare.

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References

[1] Masson C, Boukriche Y, Colombani JM. Inflammatory hypertrophic cranial pachymeningitis. Presse Med. 2001;95:65-70. Link

[2] Kupersmith MJ, Martin V, Heller G, Shah A, Mitnick HJ. Idiopathic hypertrophic pachymeningitis. Neurology. 2004;62(5):686-94. https://doi.org/10.1212/01.WNL.0000113748.53023.B7

[3] Prabhakar S, Bhatia R, Lal V, Singh P. Hypertrophic pachymeningitis: Varied manifestations of a single disease entity. Neurol India. 2002;50:45-52. Link

[4] Charcot JM, Joffroy A. Deux cas d'atrphie musculaire progressive avec lesions de la substance grise et des faisceaux anterolater- aux de la moelle epiniere. Arch Physiol Norm Pathol.1869;2:354-67 629-49, 744-6. Link

[5] Oiwa Y, Hyotani G, Kamei I, Itakura T. Idiopathic hypertrophic cranial pachymeningitis associated with total occlusion of the dural sinuses-case report. Neurol Med Chir (Tokyo). 2004;44(12):650-4. https://doi.org/10.2176/nmc.44.650

[6] Bhatia R, Tripathi M, Srivastava A, Garg A, Singh MB, Nanda A, et al. Idiopathic hypertrophic cranial pachymeningitis and dural sinus occlusion: two patients with long-term follow up. J Clin Neurosci. 2009;16(7):937-42. https://doi.org/10.1016/j.jocn.2008.08.012

[7] Chittem L, Ganti S, Rajesh A. Revisiting Eiffel- at night, in brain Neurol India. 2013;61(6):667-8. https://doi.org/10.4103/0028-3886.125288

[8] Thomas B, Thamburaj K, Kesavadas C. 'Eiffel-by-Night': A New MR Sign Demonstrating Reactivation in Idiopathic Hypertrophic Pachymeningitis. Neuroradiol J. 2007;20(2):194-5. https://doi.org/10.1177/197140090702000211

[9] Stam J. Thrombosis of the cerebral veins and sinuses. N Engl J Med. 2005;352(17):1791-8. https://doi.org/10.1056/NEJMra042354

[10] De Virgilio A, de Vincentiis M, Inghilleri M, et al. Idiopathic hypertrophic pachymeningitis: an autoimmune IgG4-related disease. Immunol Res. 2016. https://doi.org/10.26226/morressier.56e174d2d462b8028d88a670

[11] Ruiz-Sandoval JL, Bernard-Medina G, Ramos-Gomez EJ, et al. Idiopathic hypertrophic cranial pachymeningitis successfully treated with weekly subcutaneous methotrexate. Acta Neurochir (Wien). 2006;148:1011-4. https://doi.org/10.1007/s00701-006-0775-8

[12] Kleiter I, Schuierer G, Marienhagen J, et al. Intraventricular cytarabine in a case of idiopathic hypertrophic pachymeningitis. J Neurol Neurosurg Psychiatry. 2004;75:1346-8. https://doi.org/10.1136/jnnp.2003.024653

[13] Einhäupl K, Bousser MG, de Bruijn SF, et al. EFNS guideline on the treatment of cerebral venous and sinus thrombosis. Eur J Neurol. 2006;6:553-9. https://doi.org/10.1111/j.1468-1331.2006.01398.x