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Early Azathioprine as steroid-sparing therapy in Chronic Hypertrophic Pachymeningitis

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Introduction: Hypertrophic pachymeningitis (HP) is an uncommon inflammatory disease of unknown origin, characterized by abnormal focal or diffuse thickening of the dura mater. HP may arise idiopathically or secondarily from autoimmune disorders (IgG4 related disease), infections and neoplastic/infiltrative processes. Headache represents the most frequent clinical presentation, and roughly 50 % of patients relapse after corticosteroid therapy. We present two HP cases that highlight the role of azathioprine (AZA) as a maintenance therapy in the long-term management of this condition.

Case report: the first case, a 64 year old man, was referred for progressive bilateral hypoacusis and tinnitus. Brain MRI showed symmetrical supra and infratentorial dural thickening with intense enhancement involving both internal auditory canals. A complete infectious screening was negative. CSF analysis demonstrated albumino cytologic dissociation with normal IgG4 levels, supporting an idiopathic inflammatory origin.
The second case, a 52 year old man, began with headache after catarrhal otitis media, followed by deafness, dysarthria, hypophonia and rightward tongue deviation. Brain MRI demonstrated left transverse and sigmoid sinus thrombosis accompanied by left otomastoiditis and contiguous pachymeningeal enhancement. The venous thrombosis was treated while serial brain imaging showed progressive dural thickening and enhancement, suggesting an evolving inflammatory pachymeningitis. Secondary causes were repeatedly excluded through serology and CSF analysis, indicating an idiopathic form.

Discussion: the first patient received high dose iv methyl prednisolone followed by oral steroid, auditory symptoms rapidly improved, but the development of a central serous chorioretinopathy forced discontinuation. Oral azathioprine was introduced as maintenance monotherapy and, over the following 36 months, produced gradual clinical recovery together with an almost complete disappearance of pachymeningeal enhancement on MRI. The second patient experienced two flares while tapering corticosteroids, so oral Azathioprine was recently started. Since its introduction he has remained free of headache and cranial nerve deficits, and a 12 month MRI is scheduled to confirm radiological response.

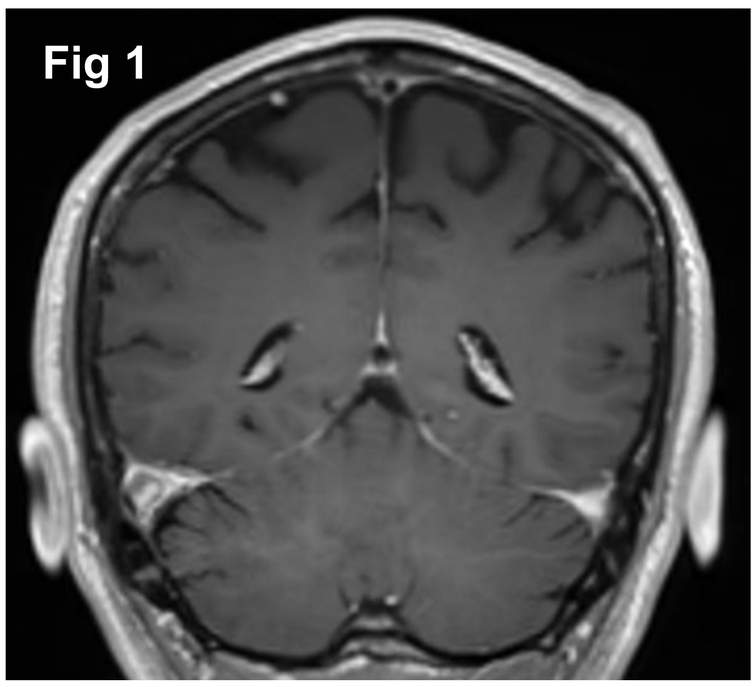

Fig.1 first patient's T1-weighted + C brain MRI showing a near-complete resolution of pachymeningeal enhancement

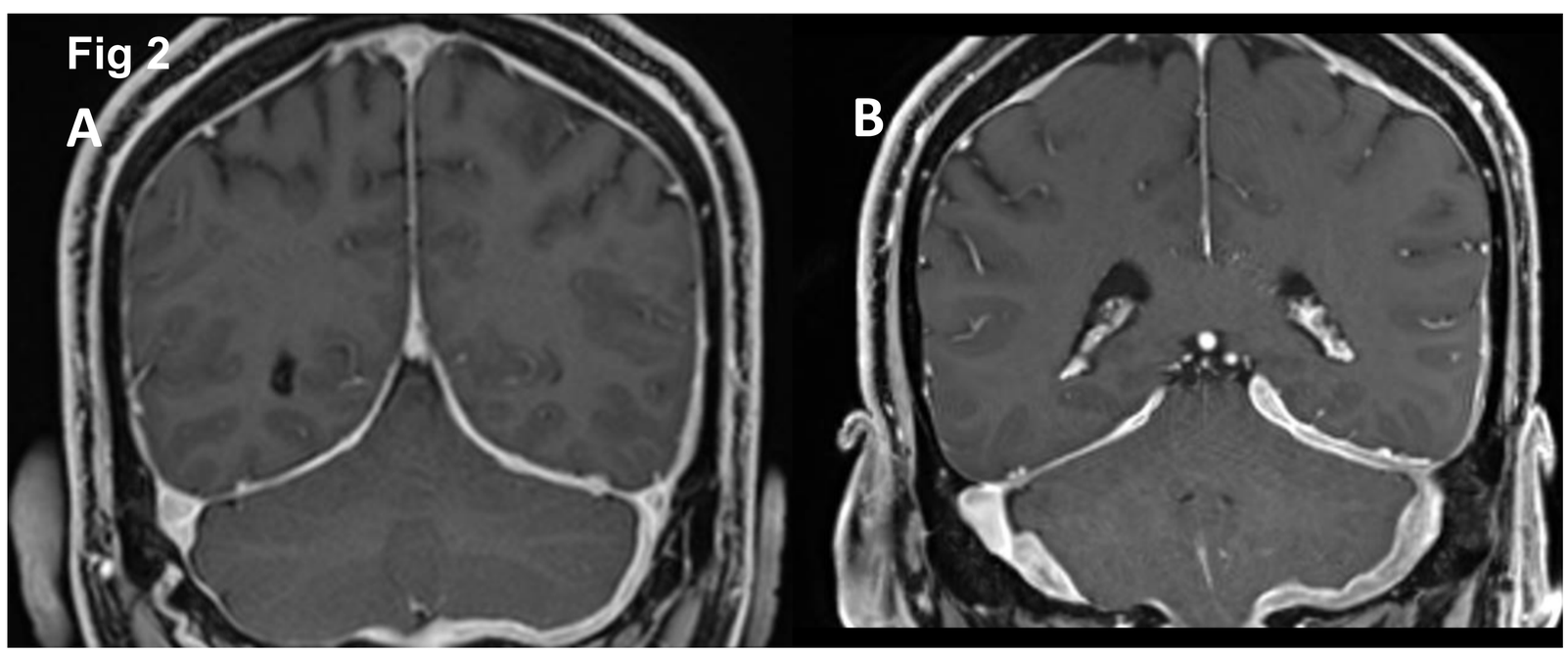

Fig.2 second patient's T1-weighted + C brain MRI in 2023 (A) vs 2025 (B) showing progressive worsening of pachymeningeal enhancement despite multiple courses of oral corticosteroid therapy

Conclusion: these two cases highlight the importance of appropriate timing regarding steroid sparing therapy introduction in idiopathic hypertrophic pachymeningitis. The first patient, switched to azathioprine after only six weeks of steroids, getting three years of uninterrupted clinical and radiological remission. The second patient, managed with steroids alone at first, suffered two definite relapses before azathioprine was added, and has been stable only since then. As reported in literature introducing azathioprine early sustains remission, reduces steroid burden and safeguards cranial nerve function. Rapid diagnosis, exclusion of secondary causes and a timely switch to azathioprine, especially where biologics are unavailable, can prevent flares and improve quality of life.

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