IgG4-related hypertrophic pachymeningitis case reports

Hypertrophic Pachymeningitis and Interstitial Lung Disease in IgG4-Related Disease [1].

A 40-year-old man presented with headache and bilateral abducent paralysis. He was also diagnosed with pulmonary tuberculosis 10 years ago and was on regular treatment for the same. Before the operation and steroid therapy, the patient was suspected of having tubercular meningitis at a local hospital. A clivus lesion was found via brain magnetic resonance imaging (MRI) at this presentation. He was preliminarily diagnosed with meningioma and underwent Gamma Knife Surgery. Transnasal endoscopic resection was performed to treat deterioration of nerve function. Postoperative pathologic examination suggested IgG4-RD. Moreover, the serum IgG4 was elevated at 1.90 g/L (reference range: 0.035-1.500 g/L). After steroid therapy for 2 mo, the lesion size diminished on MRI, and the function of bilateral abducent nerves recovered.

IgG4-RHP is relatively rare and indistinguishable before the operation. Elevated serum IgG4 levels and imaging examination help in the diagnosis of IgG4-RHP. Surgery is necessary when lesions progress and patients start to develop cranial nerve function deficit [2].

A case of a young-man presenting with thoracic spinal epidural compressive lesion which was indeed a manifestation of IgG4-related hypertrophic pachymeningitis. The mass was firmly adherent to the dura and extended into left neural foramen/paravertebral space which precluded complete excision. Frozen sections suggested fibro-inflammatory stroma with large areas of fibrosis and lymphoplasmacytic infiltrate. After subtotal excision, the patient improved with medical therapy at 1-year follow-up. Although uncommon, the case highlights the need to consider spinal presentation of this rare entity, especially in the context of autoimmune disorders or even in isolation. In this regard, intraoperative frozen section can hint the underlying inflammatory/autoimmune pathology, guide further course of surgery as well as limit unwarranted operative morbidity [3].

A 35-year-old woman presented with a 6-month history of neck and right shoulder pain, progressive right triceps weakness and paresthesias in the right C8 and T1 dermatomes. MRI demonstrated a T2 hypointense epidural soft tissue mass extending from C6-T1. The patient underwent C6-T1 laminoforaminotomy and partial resection with near complete symptom resolution. Histopathology was consistent with diagnosis of IgG4-RHP. Eighteen months postoperatively, she experienced symptom recurrence necessitating re-operation and adjuvant postoperative prednisone with complete resolution at 40-months' follow-up.

Of the now nineteen confirmed cases of IgG4-RHP, fifteen underwent surgery. A majority achieved partial resection. Three surgical patients did not receive adjuvant therapy with symptomatic recurrence between 2 and 18 months [4].

A 68-year-old man presented at a nearby hospital with a headache and a low-grade fever. A blood test revealed inflammation, as well as elevation of IgG4 level. Magnetic resonance imaging(MRI)revealed diffuse thickening of the dura mater, dominantly in the posterior fossa and cerebellar tentorium. The lesion was enhanced significantly with gadolinium(Gd). An open biopsy was performed to determine pathological diagnosis. Hematoxylin and eosin staining showed infiltration of inflammatory cells, including plasma cells. The infiltrating cells were positive for IgG4. Post-operatively, the patient was treated with glucocorticoid, and both the inflammation and patient symptoms were improved. In conclusion, IgG4 is related to the etiology of hypertrophic pachymeningitis and glucocorticoid therapy is effective for this disease [5)].

Takeuchi et al., report the tenth case of IgG4-related intracranial hypertrophic pachymeningitis and review the previous literature. A 45-year-old male presented with left-sided focal seizures with generalization. Magnetic resonance imaging (MRI) revealed a diffuse thickening and enhancement of the right convexity dura matter and falx with focal nodularity. The surgically resected specimens exhibited the proliferation of fibroblast-like spindle cells and an infiltration of mononuclear cells, including predominantly plasma cells. The ratio of IgG4-positive plasma cells to the overall IgG-positive cells was 45% in the area containing the highest infiltration of plasma cells. On the basis of the above findings, IgG4-related sclerosing disease arising from the dura mater was suspected. IgG4-related sclerosing disease should be added to the pachymeningitis spectrum [6)].

Popkirov et al., describe a 52-year-old man with hypertrophic pachymeningitis (HP) who was both seropositive for antineutrophil cytoplasmatic antibodies (ANCA) against myeloperoxidase, and had an immunoglobulin G4 (IgG4) positive fibroinflammatory response in meningeal biopsy. HP is a chronic inflammatory thickening of the dura mater which typically presents with headache, cranial nerve dysfunction and other neurological deficits. While first-line treatment with corticosteroids is recommended, many patients relapse and need additional immunosuppression. One recently described etiology is IgG4-related disease and in a subgroup of idiopathic patients, evidence suggests a crucial role of ANCA. To our knowledge, the simultaneous occurrence of IgG4-related disease and ANCA has not been reported so far. This man suffered life-threatening disease progression despite the administration of high dose steroids, cyclophosphamide and azathioprine. Treatment with Rituximab was initiated which led to disappearance of clinical symptoms and decrease of dural thickening within weeks. This patient presents a possible disease overlap of IgG4-related and ANCA-associated HP and illustrates the effectiveness of Rituximab in refractory IgG4-related HP [7)].

1)
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