CASE REPORT

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Rare Presentation of CNS Manifestation of Pseudo-Inflammatory Tumor which Responded to Steroid: A Case Report

ABSTRACT

Aim: Rare presentation of central nervous system manifestation of pseudo-inflammatory tumor which responded to steroids. **Background:** Inflammatory pseudotumors are rare, benign, tumor-like lesions characterized by chronic inflammatory cell infiltration, which can mimic malignant tumors both clinically and radiologically. **Case Description:** Here, we present a case of a patient diagnosed with an inflammatory pseudotumor involving the left 7th and 8th cranial nerves, who presented with symptoms of severe headache, recurrent vomiting, dizziness, and gait ataxia. Initial examination revealed no fever or other systemic symptoms, and magnetic resonance imaging showed enhancement along the left 7th and 8th nerve complex. **Conclusion:** Elevated serum immunoglobulin G4 levels supported an inflammatory etiology, whereas cerebrospinal fluid analysis and other tests ruled out infection or malignancy. The patient showed significant improvement in symptoms after corticosteroid therapy with IV dexamethasone and supportive care. **Clinical Significance:** This case highlights the diagnostic challenge posed by inflammatory pseudotumor in cranial nerves and underscores the importance of distinguishing them from other lesions to guide appropriate treatment.

Key words: 7th and 8th nerve involvement, Benign lesion, Immunoglobulin G4, Magnetic resonance imaging skull base

INTRODUCTION

The inflammatory pseudotumor was coined in 1954. Inflammatory pseudotumors are rare benign lesions characterized by chronic inflammatory cell infiltration. They may mimic malignant tumors both radiologically and clinically.[1] It is a diagnosis of exclusion after ruling out lymphomas, sarcomas, or sarcoidosis.[2] The differential diagnosis includes such as neoplastic (lymphomas, sarcomas, and carcinomas in general), immunological (sarcoidosis, Wegener's granulomatosis, and vasculitis, immunoglobulin G4 [IgG4] disease), or infectious (such as tuberculosis, aspergillosis, or mucormycosis, among others) pathologies.[3] Histopathological findings include those processes in which an abundant inflammatory infiltrate of lymphoplasmacytic predominance and spindle cells with myofibroblast phenotype without atypia, along with the formation of lymphoid follicles, is observed in the biopsy of the affected organ.[4]

CASE REPORT

A 52-year-old female patient presented with headache, vomiting, and dizziness. She also complained of gait ataxia with a tendency to fall toward the left side for the last 3 days. The headache was throbbing, generalized, and associated with non-bilious vomiting. Headache increases upon waking and is present throughout the day. There was no fever or convulsion. She also developed left facial weakness with pain in her left eye for 2 days. She does not have any comorbidities. On examination, she was hemodynamically stable. Afebrile, vital parameters are normal. Central nervous system examination revealed consciousness, orientation, and all higher functions were normal. She was having left lower motor neuron facial weakness. No other cranial nerve involvement. Her cerebellar system examination revealed mild gait ataxia with a tendency to fall toward the left side and superficial and deep reflexes were normal. Plantars are down going. Neither neck stiffness nor meningeal signs are present.

Her investigations showed Hb. 10.7/7600/198k. Her electrolytes were Na 138, K 4.3, and CL 110. Her C-reactive protein 87, and PCT 0.05, Liver Function tests, Urine routine, se. Vit D and se. Vit B12 was within normal limits. Her coagulation profile BT/CT/PT/INR was also normal. Creatine 0.9, triple H – negative.

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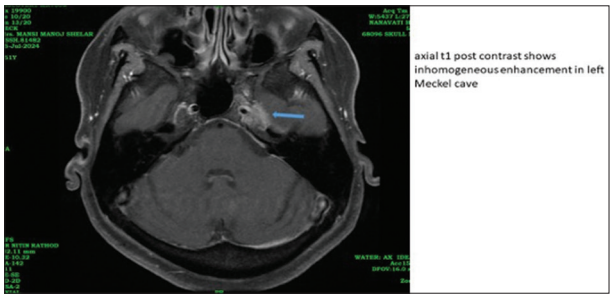

Figure 1:

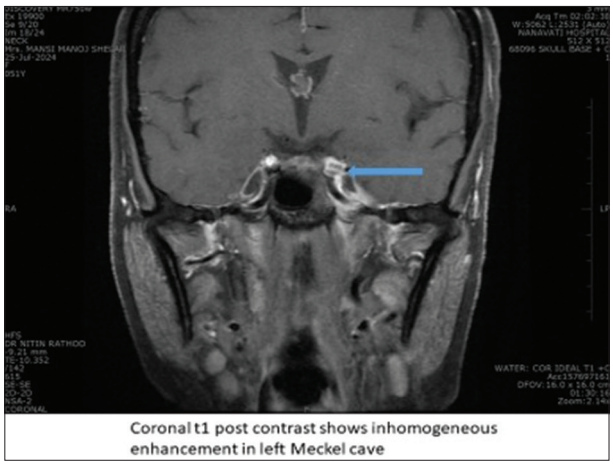

Figure 2:

Cerebrospinal fluid (CSF) examinations revealed: Clear, colorless, 4 cells/cu mm, all are lymphocytes. CSF protein – 40 mg/dL, CSF sugar – 60 mg/dL, CSF culture – no growth, CSF smear – no pus cells, no organisms, no fungal, and no AFB bacilli are seen. In addition, the smear did not show malignant cells. CSF – meningoencephalitis bio fire (viruses, bacteria, and yeast) – negative. CSF adenosine deaminase activity – O.56 U/L (normal 0–30). CSF angiotensin-converting enzymes level 2.0 U/L. CSF IG G4 level was 0.3 G/L (more than 1.4 G/L is significant). CSF bacterial and fungal culture – no growth, CSF – cryptococcus Ag – negative.

Magnetic resonance imaging (MRI) of the brain with MRI skull base with contrast study revealed inhomogeneous enhancement involving the 7th and 8th nerve complex, left mackle's cave, left cavernous sinus along the wall of petrous and adjoining the vertical segment of the internal carotid artery, left superolateral aspect of nasopharynx on the left side and patchy meninges on the above-described structures. The rest of the brain parenchyma and meninges are normal [Figures 1 and 2]. Positron emission tomography scan: No abnormalities detected in the entire body. Since the patient was having a headache, vomiting, left LMN facial weakness, and left-sided mild cerebellar ataxia, we were thinking of intracranial pathology. MRI/magnetic resonance angiography of the brain was normal. MRI skull base with contrast revealed

Figure 3:

enhancement of left 7th and 8th cranial nerve affection with the surrounding area having a differential diagnosis of (1) inflammatory pseudotumor, (2) IGG4-related inflammatory response, and (3) lymphoma. However, since CSF IGG4 was normal and lymphoma was ruled out. After extensive investigations and diagnosis of an inflammatory tumor, she was given a steroid high dose, and the patient improved within 4–5 days with tapering dose, and she was discharged. After 7 days, no headache, no vomiting, facial weakness improved, and no gait ataxia at all.

DISCUSSION

Inflammatory pseudotumors, also known as inflammatory myofibroblast tumors, are rare benign lesions, often of idiopathic origin, that can mimic neoplastic processes due to their mass-like appearance on imaging and presentation with localized symptoms. Although they commonly involve the lung, liver, or gastrointestinal tract, their occurrence in cranial nerves is rare. The involvement of the left 7th (facial) and 8th (vestibulocochlear) cranial nerves, as seen in this patient, is particularly unusual [Figure 3]. Symptoms such as headache, vomiting, dizziness, and gait ataxia can resemble those of other cranial nerve pathologies, such as neoplastic, vascular, or infectious processes, making diagnosis challenging. The mainstay of treatment is corticosteroids. As in systemic diseases, some authors propose the use of immunosuppressants to reduce the doses, duration, and complications of corticosteroids.[5]

CONCLUSION

This case report highlights the successful diagnosis and management of an inflammatory pseudotumor involving the left 7th and 8th cranial nerves, presenting neurological symptoms that initially raised concern for malignancy or other

severe pathology. The combination of MRI findings elevated IgG4 levels, and a positive response to corticosteroids supported the diagnosis and helped differentiate it from more aggressive pathologies. This case emphasizes the value of including inflammatory pseudotumor in differential diagnoses of cranial nerve lesions and suggests that corticosteroid therapy can lead to effective symptom management in these patients. Follow-up care, including outpatient monitoring, will be essential for detecting any recurrence and ensuring long-term stability.

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