International Journal of Science and Research (IJSR)
ISSN: 2319-7064
ResearchGate Impact Factor (2018): 0.28 | SJIF (2018): 7.426

Neurosarcoidosis - Rare Presentation with Multiple Cranial Nerve Involvement

Dr Anand Somkuwar[1], Dr Archit Gupta[2]

[1]DM, Neurology, Topiwala National Medical College& B.Y.L Nair Ch Hospital, Mumbai, India

[2]MD Radiodiagnosis, Topiwala National Medical College& B.Y.L Nair Ch hospital, Mumbai, India

Abstract: ***We report a rare case of neurosarcoidosis with multiple cranial nerve involvement in the absence of pulmonary features. It is estimated that less than 1% of patients have an isolated CNS involvement, without systemic evidence of disease. A 42 year old man presented with symptoms of multiplecranial neuropathy. His MRI Brain showed abnormal enhancement in left trigeminal and facial nerve with bulky bilateral lacrimal glands. HRCT Thorax demonstrates perilymphatic nodules with patchy ground glass opacities and non-necrotic mediastinal lymphadenopathy. Patient underwent CT guided mediastinal lymph node biopsy which revealed non-caseating chronic granuloma, consistent with sarcoidosis.***

Keywords: Neurosarcoidosis, Multiple Cranial nerve, lacrimal gland, Magnetic Resonance Imaging

1. Case Report

A 42 years old male admitted with complaint of pain and numbness on the left side of face, decreased taste sensation on left side of tongue with deviation of angle of mouth to right side.

No history of fever/decreased vision/diplopia/nasal regurgitation/focal neurodeficit.Patient was non-diabetic,non-hypertensive,non-alcoholic.

On CNS examination-Patient has left ophthalmic, maxillary and mandibular nerve involvement with left facial nerve involvement.

No tympanic membrane perforation, no mastoid tenderness, no neck stiffness.

Patient undergone MRI Brain which reveals abnormally thickened and enhancing cisternal segment of left trigeminal nerve and mandibular division in its proximal segment(Figure 1,2,3,4).Mild enhancement is seen along the horizontal and vertical segment of left facial nerve(Figure5).Bilateral bulky and abnormally enhancing lacrimal glands(Figure 6,7).

No signs of meningeal enhancement are seen.Pituitary appears normal in size and enhancement.Patient undergone CSF examination which shows normal cellularity and no malignant cells, also CSF was negative for Mycobacterium Tuberculosis.
HRCT Thorax revealed multipleperilymphatic nodules with patchy ground glass opacities was seen and non-necrotic mediastinal lymphadenopathy. Patient further underwent transbronchial lung biopsy which confirmed the diagnosis of sarcoidosis.

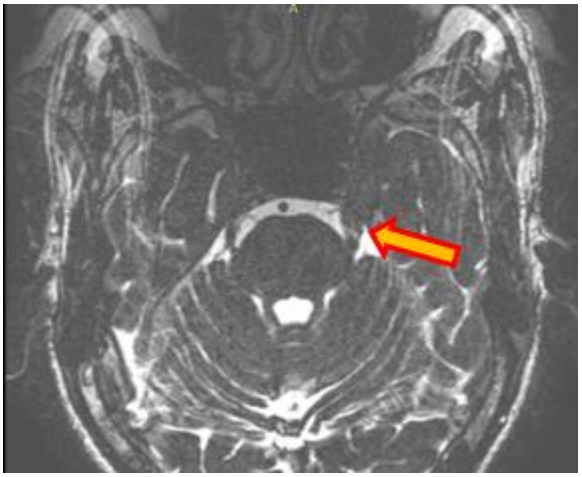

Figure 1: T2 VISTA AXIAL

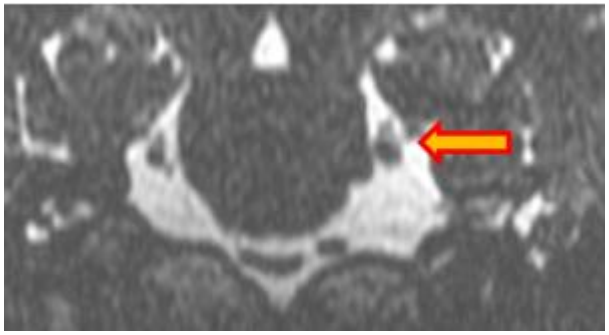

Figure 2: T2 VISTA

CORONALT2 VISTA images clearly demonstrating bulkyl left trigeminal nerve

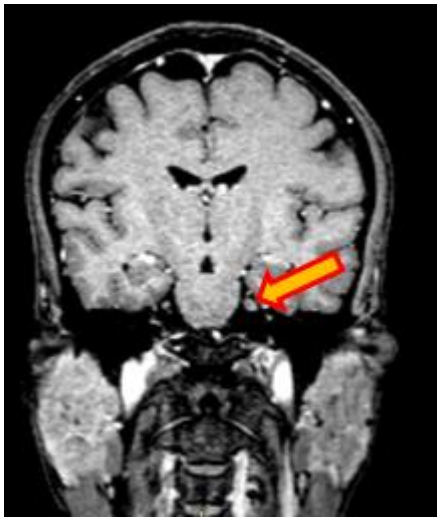

Figure 3: T1 PC CORONAL

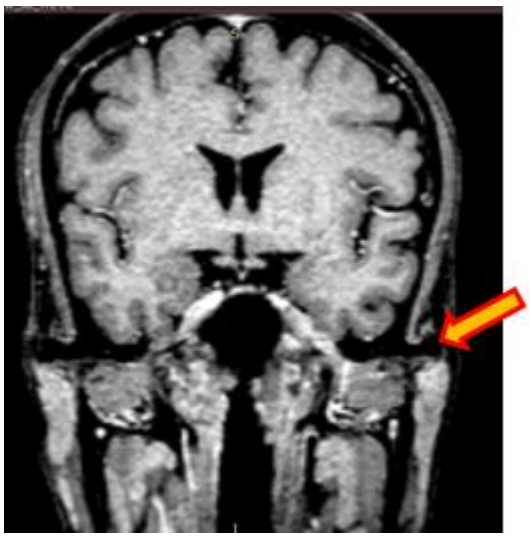

Figure 4: T1 PC CORONAL

T1 PC images showing abnormally enhancing left trigeminal nerve and its mandibular division upto foramen ovale as shown in Figure 4.

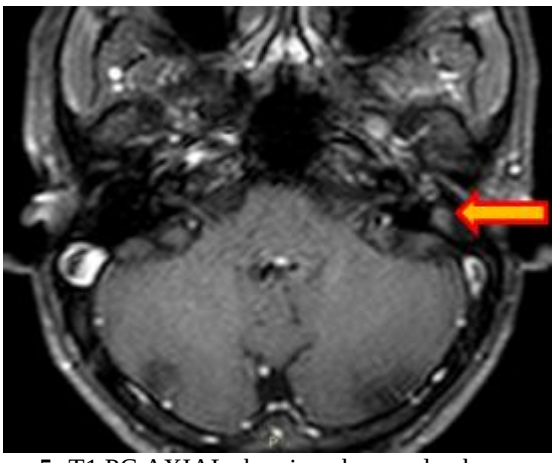

Figure 5: T1 PC AXIAL showing abnormal enhancemnt in left facial nerve

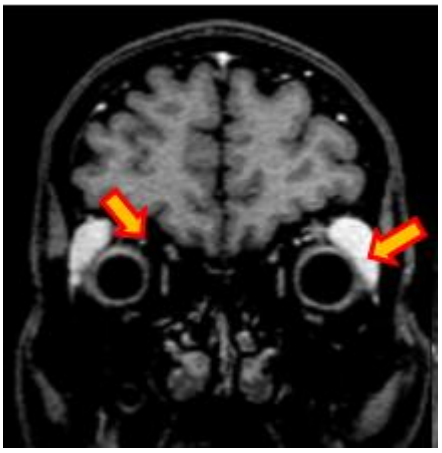

Figure 6: T1 PC CORONAL

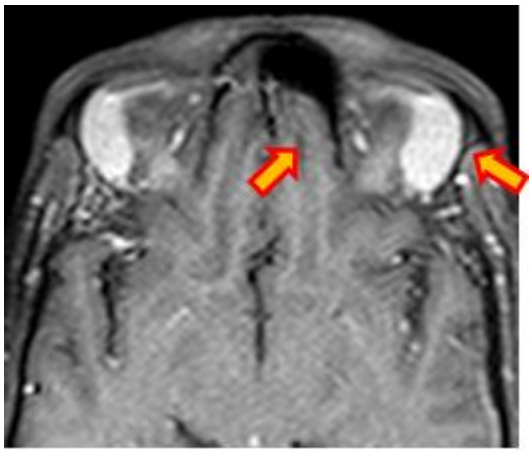

Figure 7: T1 PC AXIAL

Coronal and axial sections demonstrating bilaterally bulky lacrimal glands.

2. Discussion

Neurosarcoidosis is a rare variant of sarcoidosis, which can have varying presentations, including headaches, seizures, ataxia, visual disturbances, loss of sensation, and other neurologic abnormalities. Although most cases of sarcoidosis are treatable, neurosarcoidosis can have mortality as high as 10%. Imaging studies and biopsy are helpful in confirming the diagnosis. Corticosteroids, immunomodulators, and close follow-up are critical in managing the symptoms and reducing long-term sequelae of the condition. Our patient improved symptomatically with steroids and immunosuppressant.

The diagnosis of neurosarcoidosis can be difficult due to the variability ofpatient'spresentation which includes cranial nerve palsies, meningitis, seizures, andneuropsychiatric symptoms [1, 2].

Multiple cranial neuropathies are usually uncommon presentations with diverse causes. Tumourappears to be the commonest cause for multiple cranial neuropathies accounting for about 30% of all the cases. This isfollowed by other causes like vascular disease, trauma, and infection.

In Keane's review of 979 cases with multiple cranial palsies, sarcoidosis was the cause in 2 patients only [3].The most

common neurologic manifestation of sarcoidosis is cranial neuropathy likely secondary to nerve granulomas, raised intracranial pressure, or granulomatous meningitis [4].

The facial nerve is the most frequently affected cranial nerve in sarcoidosis followed by optic nerve. Other cranial nerves are less frequently affected.

Although the cause of sarcoidosis is unknown, an abnormal immune response toinfectious and non-infectious environmental factors appears to be a key causal factor.

Thediagnosis is confirmed based on clinical and radiological correlation in combination with a tissuebiopsy [4, 5].

Although commonly requested, serum angiotensin converting enzyme (ACE) has a poor predictive value in diagnosing sarcoidosis. In our patient the serum ACE level was normal.

A normal MRI Brain does not rule out the possibility ofneurosarcoidosis. A chest X-ray is helpful, as it can display mediastinal lymphadenopathy as was the case in our patient.

References

[1] K. Nozaki and M. A. Judson, “Neurosarcoidosis: clinical manifestations,diagnosis and treatment,” La PresseMédicale, vol. 41, no. 6, part 2, pp.e331–e348, 2012. View at Publisher · View at Google Scholar
[2] E. Hoitsma, C. G. Faber, M. Drent, and O. P. Sharma, “Neurosarcoidosis: aclinical dilemma,” The Lancet Neurology, vol. 3, no. 7, pp. 397–407, 2004. Viewat Publisher · View at Google Scholar · View at Scopus
[3] J. R. Keane, “Multiple cranial nerve palsies: analysis of 979 cases,” Archives ofNeurology, vol. 62, no. 11, pp. 1714–1717, 2005. View at Publisher · View atGoogle Scholar · View at Scopus
[4] S. Pawate, H. Moses, and S. Sriram, “Presentations and outcomes ofneurosarcoidosis: a study of 54 cases,” Quarterly Journal of Medicine, vol. 102,no. 7, pp. 449–460, 2009. View at Publisher · View at Google Scholar · View atScopus
[5] Seltzer S, Mark AS, Atlas SW. CNS sarcoidosis: evaluation with contrast enhanced MR imaging. AJNR Am J Neuroradiol 1991; 12: 1227-33.