Multiple cranial nerves involvement as clinical onset of neurosarcoidosis

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INTRODUCTION

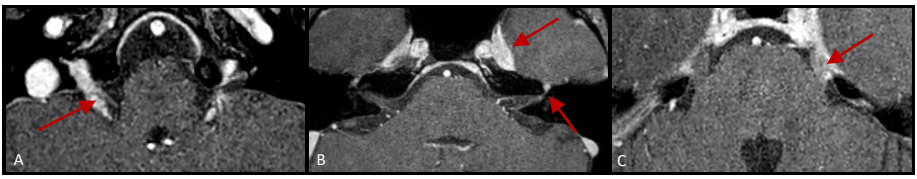
Multiple cranial neuropathies (MCN) present a diagnostic challenge due to a broad spectrum of possible etiologies, including autoimmune, vascular, infectious, neoplastic, and traumatic causes. We describe a patient who presented with left trigeminal nerve impairment, progressive dyspnea, and dysphagia due to multiple cranial nerves involvement. Radiological findings and mediastinal lymphadenopathy raised suspicion for cranial neurolymphomatosis, but further investigation led to diagnosis of neurosarcoidosis. Neurolymphomatosis, caused by malignant lymphomatous cells infiltration, often present with multifocal neuropathies.
In contrast, neurosarcoidosis, which affects 5-10% of sarcoidosis patients, usually presents at disease onset or within the first two years with cranial neuropathies (50-70% of cases) and presents as monophasic, relapsing-remitting, or progressive disease.

MATERIALS AND METHODS

A 48-year-old man presented with progressive paresthesia starting in the right lower lip, extending over two months to the left side and further to the left zygomatic and temporal regions. He also developed complete dysphagia.
Diagnostic workup included ear, nose, and throat (ENT) assessment, videolaryngoscopy, neck ultrasound, brain CT and MRI with contrast, whole-body PET/CT, blood tests, lumbar puncture, endoscopic biopsy with endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) and bronchoalveolar lavage (BAL).

RESULTS

Neurological evaluation revealed multiple cranial nerve deficits including hypophonic speech, right hemipalatal and laryngeal paralysis. Left facial nerve palsy developed during hospitalization. Blood tests showed elevated PCR, soluble interleukin-2 receptor, and ACE. Cerebrospinal fluid analysis revealed increased protein (76 mg/dL) and mild pleocytosis (13 lymphocytes/µL). Brain MRI showed enhancement of the left V and VIII nerves and the right IX, X, XI nerves. PET/CT revealed intense hypermetabolism in mediastinal lymphadenopathy. At BAL, increased lymphocytes and a markedly elevated CD4/CD8 ratio was present. EBUS-TBNA revealed granulomatous, non-necrotizing formations. The patient was diagnosed with neurosarcoidosis and treated with high-dose corticosteroids, methotrexate, and infliximab with complete resolution of symptoms. A slight tiny paresthesia on the left cheek persists. Follow-up MRIs showed significant reduction in cranial nerve enhancement (1-month follow-up), with complete resolution at the 3- and 4-year follow-up.



A) Hyperintensity and swelling of the right cranial nerves IX, X, and XI. B) Meckel's caves filled with enhancing tissue (complete on the left, nearly complete on the right). Subtle enhancement in the left internal auditory canal along the course of the vestibulocochlear nerve (VIII).
C) Enhancement involving the remaining segments of the left trigeminal nerve (pre-pontine cisternal portion and foramen ovale).

CONCLUSION

This case underscores the diagnostic challenges posed by multiple cranial neuropathies, where the differential diagnosis may encompass rare conditions such as neurosarcoidosis and neurolymphomatosis. The lack of specific biomarkers in both conditions complicates the diagnostic workup, making histopathological examination essential.
A multidisciplinary approach, integrating clinical, radiological, and laboratory data, is crucial for accurate diagnosis and management. Regular clinical and radiological follow-up is essential to assess disease progression and therapy response, optimizing outcomes and minimizing complications.