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NEUROSARCOIDOSIS WITH EXTENSIVE OPTO-CHIASMATIC AND MENINGEAL INVOLVEMENT: A CASE REPORT

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AUTHOR AND AFFILIATION

A. Bijbij[1], L. Belkouchi[1], Zhim Meriem[1], S. El Haddad[1], N. Allali[1], L. Chat[1]

[1]Service d'Imagerie Femme-Enfant, Hôpital des Enfants de Rabat, CHU Ibn Sina, Rabat, Morocco

Corresponding author: Ayman Bijbij, MD

ABSTRACT

Neurosarcoidosis, a rare neurological manifestation of systemic sarcoidosis, poses significant diagnostic challenges due to its nonspecific clinical presentation and variable imaging features. We report the case of a 64-year-old female presenting with progressive visual disturbances and multifocal neurological deficits. Magnetic resonance imaging (MRI) revealed extensive opto-chiasmatic involvement, diffuse nodular leptomeningeal enhancement, and cranial nerve inflammation, consistent with neurosarcoidosis. This case underscores the indispensable role of advanced neuroimaging in delineating disease extent, guiding biopsy, and monitoring therapeutic response. Early recognition of characteristic MRI patterns, combined with multidisciplinary collaboration, is critical to prevent irreversible neurological damage and optimize patient outcomes.

KEYWORDS

Neurosarcoidosis, Leptomeningeal enhancement, MRI

MAIN ARTICLE

Introduction

Sarcoidosis is a multisystem granulomatous disorder of unknown etiology, predominantly affecting the lungs and lymphatic system. Neurological involvement, termed neurosarcoidosis, occurs in fewer than 5% of cases but carries substantial morbidity due to its propensity to involve critical structures such as the optic pathways, meninges, and cranial nerves. Diagnosis is often delayed due to the lack of pathognomonic biomarkers and the rarity of histopathological confirmation from neural tissue. Neuroimaging, particularly contrast-enhanced MRI, serves as the cornerstone of evaluation, enabling visualization of granulomatous inflammation and exclusion of mimics such as infections or malignancies. This report highlights a complex case of neurosarcoidosis with extensive opto-chiasmatic and leptomeningeal involvement, emphasizing the synergy between imaging and clinical decision-making.

Case Report :

Clinical Presentation

A 64-year-old female presented with a 6-month history of progressive bilateral visual blurring, right-sided facial numbness, and intermittent headaches. Neurological examination revealed decreased visual acuity (20/60 in the right eye, 20/40 in the left) and hypoesthesia in the right trigeminal nerve (V1–V2 distribution). Serum angiotensin-converting enzyme (ACE) levels were elevated at 68 U/L (normal: 12–65 U/L). Chest X-ray demonstrated bilateral hilar lymphadenopathy, prompting further evaluation with brain and spinal MRI.

Imaging Findings

A comprehensive MRI protocol was performed, including T1-weighted, T2-weighted, FLAIR, diffusion-weighted imaging (DWI), and post-gadolinium 3D T1 sequences. Key findings included:

1. **Optic Pathway Involvement**:
    - Prechiasmatic segments of both optic nerves exhibited T2 hyperintensity and marked post-contrast enhancement, more pronounced on the right (Figure 1).
    - The optic chiasm appeared spared.
2. **Leptomeningeal Disease**:

- Nodular leptomeningeal enhancement was observed in the basifrontal region (Figure2A), pontine cistern (Figure 2B), hypothalamic area, and along the superior cerebellar peduncle.
- Additional involvement of the posterior cervical spinal cord leptomeninges was noted (Figure 2C).

3. **Cranial Nerve Inflammation**:
   - Enhancement of the right trigeminal (CN V) (Figure3) , abducens (CN VI), and hypoglossal (CN XII) nerves.
4. **Incidental Findings**:
   - A right frontal extra-axial lesion (14 × 17 mm) with homogeneous enhancement, suggestive of a meningioma.
   - Right maxillary sinusitis with mucosal thickening.

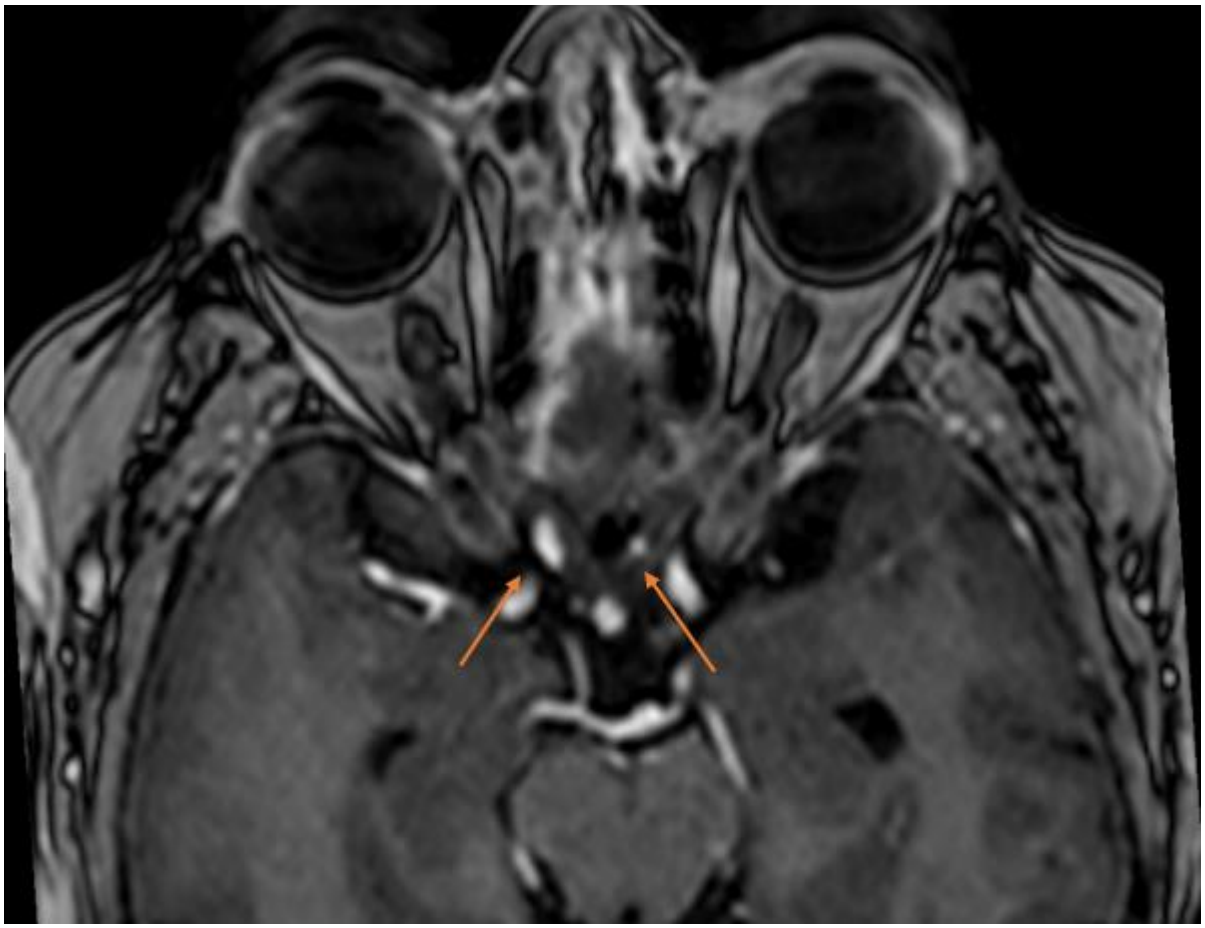
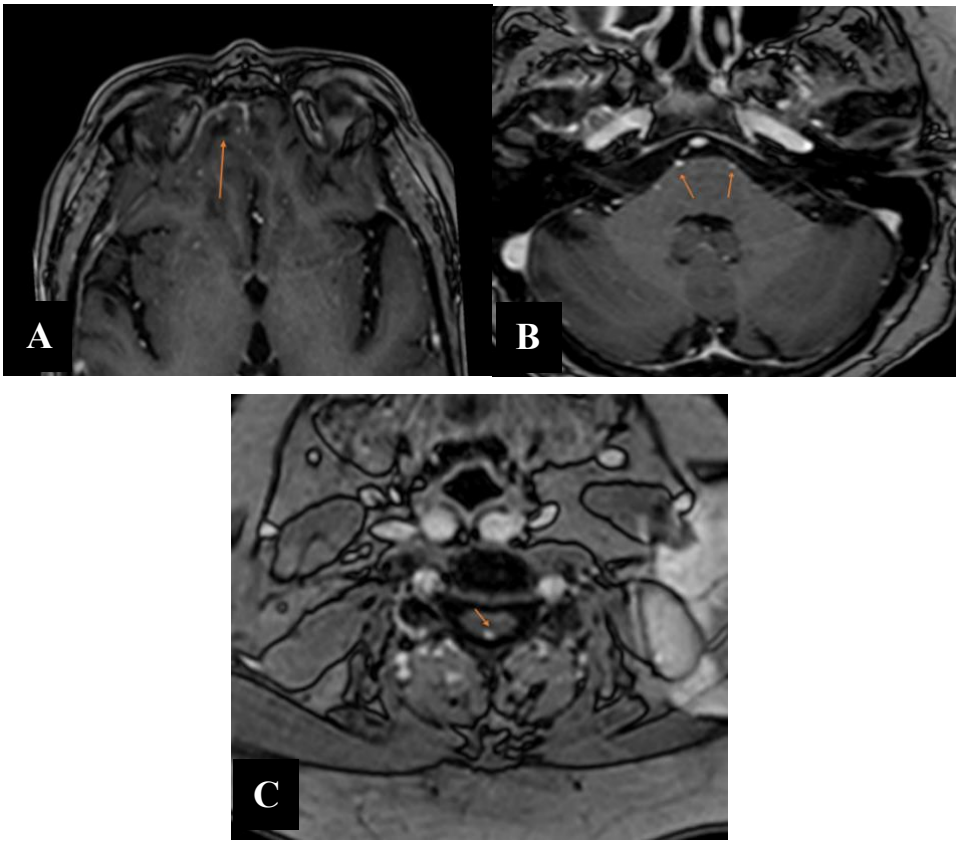
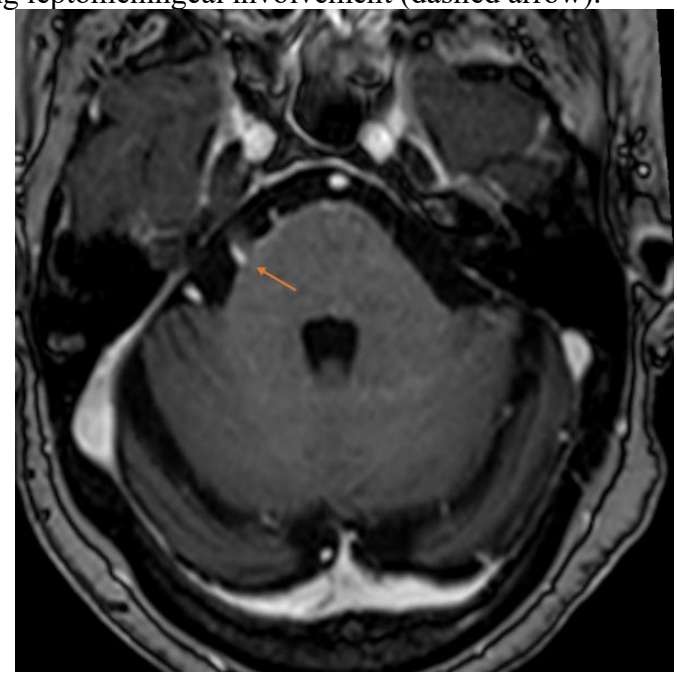


Figure 1: Post-contrast axial T1-weighted MRI demonstrating bilateral prechiasmatic optic nerve enhancement (arrows).



- **Figure 2A**: Axial post-contrast T1-weighted MRI showing nodular leptomeningeal enhancement in the basifrontal region (arrowheads).
- **Figure 2B**: Axial post-contrast T1-weighted MRI showing nodular leptomeningeal enhancement in the prepontine region (arrowheads).
- **Figure 2C**: Axial post-contrast T1-weighted MRI at the level of the posterior cervical cord, highlighting leptomeningeal involvement (dashed arrow).



- **Figure 3**: Post-contrast axial T1-weighted MRI demonstrating CN V nerve enhancement (arrows).

Histopathological Correlation

Transbronchial lung biopsy revealed non-caseating granulomas with multinucleated giant cells, confirming systemic sarcoidosis. Cerebrospinal fluid (CSF) analysis demonstrated lymphocytic pleocytosis (32 cells/μL) and elevated protein (68 mg/dL), supporting inflammatory meningeal involvement.

Management and Follow-Up

The patient was initiated on high-dose intravenous methylprednisolone (1 g/day for 5 days), followed by oral prednisone (60 mg/day). At 3-month follow-up, visual acuity improved to 20/30 bilaterally, and facial numbness resolved. Repeat MRI showed partial regression of leptomeningeal enhancement. Long-term immunosuppression with methotrexate was planned to prevent relapse.

Discussion

Neurosarcoidosis remains a diagnostic enigma due to its heterogeneous clinical and radiological presentation. In this case, MRI played a pivotal role in identifying hallmark features of the disease, including opto-chiasmatic inflammation and nodular leptomeningeal enhancement. The latter, characterized by granular or linear post-contrast uptake along the pial surface, is highly suggestive of granulomatous infiltration and distinguishes neurosarcoidosis from infectious etiologies such as tuberculosis, which typically exhibits basal exudates.

The bilateral optic nerve involvement observed here is a recognized feature of neurosarcoidosis, often mimicking optic neuritis. However, the absence of diffusion restriction and concurrent meningeal abnormalities favored a granulomatous etiology. Cranial nerve enhancement, particularly of the trigeminal and abducens nerves, further corroborated the diagnosis, as these structures are frequently involved due to their extensive meningeal coverage.

A critical differential consideration is primary CNS lymphoma, which may present with homogeneous enhancing lesions but typically lacks the multifocal leptomeningeal pattern seen here. Wegener's granulomatosis, another granulomatous disorder, was excluded due to the absence of sinonasal involvement and negative anti-neutrophil cytoplasmic antibodies (ANCA).

Clinical Implications

1. **Early Imaging**: MRI should be expedited in patients with visual deficits and systemic sarcoidosis to assess optic pathway and meningeal involvement.

2. **Biopsy Guidance**: In cases where neural biopsy is deemed high-risk, extrapulmonary tissue sampling (e.g., lung, lymph nodes) may suffice for diagnosis.
3. **Therapeutic Monitoring**: Serial MRI enables assessment of treatment response, particularly in steroid-dependent cases.

Conclusion

This case illustrates the critical role of MRI in diagnosing and managing neurosarcoidosis, particularly in patients with atypical neurological presentations. The integration of advanced imaging, histopathological correlation, and multidisciplinary collaboration ensures timely intervention, mitigating the risk of permanent neurological sequelae.

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Ethics approval and consent to participate
Ethical approval was waived by the institutional review board as this retrospective case report did not involve interventional research.

Consent for publication
Written informed consent was obtained from the patient's legal guardian for publication of this case report and accompanying images. A copy of the consent form is available for review by the Editor-in-Chief.

Availability of data and materials
No datasets were generated or analyzed for this study. Data sharing is not applicable.

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Authors' contributions

- A.B. and L.B.: Manuscript drafting, case analysis, and imaging interpretation.
- S.E.H.: Critical revision and intellectual input.
- N.A. and L.C.: Study supervision and final approval.

All authors read and approved the final manuscript and attest to the accuracy of the data.
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