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Case Report / Olgu sunumu

# Multisystem Sarcoidosis with Central Nervous System and Extrapulmonary Involvement Presenting as Facial Nerve Palsy

## Fasiyal Sinir Paralizisi ile Başvuran Multisistemik Sarkoidoz: Santral Sinir Sistemi ve Ekstrapulmoner Tutulum

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## Abstract

Sarcoidosis is a multisystem granulomatous disease of unknown etiology that most commonly affects the lungs and lymphatic system, while central nervous system involvement is rare and diagnostically challenging. We report a case of multisystem sarcoidosis presenting with facial nerve palsy and a solitary enhancing brain lesion. A 41-year-old woman presented with right-sided facial paralysis. Contrast-enhanced brain MRI revealed right lateral ventricular enlargement with irregular walls, pituitary gland thickening and enhancement, a 6 mm enhancing nodular lesion in the left parietal lobe, and bilateral lacrimal gland enlargement. Axillary lymph node biopsy demonstrated granulomatous inflammation. Thoracic CT showed lymphadenopathy, perilymphatic nodules, and lytic rib lesions indicating osseous involvement. Elevated ACE levels supported the diagnosis. Based on clinical, radiological, and histopathological findings, multisystem sarcoidosis with neurosarcoidosis was diagnosed. This case highlights the importance of considering sarcoidosis in patients with solitary brain lesions and cranial neuropathies, especially in the presence of systemic involvement.

**Keywords**: Sarcoidosis, neurosarcoidosis, multisystem disease

## Öz

Sarkoidoz, nedeni bilinmeyen, çoklu sistemleri etkileyen granülomatöz bir hastalıktır ve en sık akciğerleri ile lenfatik sistemi tutar. Merkezi sinir sistemi tutulumu nadirdir ve tanı koymak zordur. Bu çalışmada, yüz siniri felci ve beyinde tek bir kontrast tutan lezyon ile ortaya çıkan multisistemik sarkoidoz vakası sunulmuştur. 41 yaşında bir kadın hasta sağ taraflı yüz felci ile başvurdu. Kontrastlı beyin MRG'sinde sağ lateral ventrikülde düzensiz duvarlı genişleme, hipofiz bezinde kalınlaşma ve kontrast tutulumu, sol parietal lobda 6 mm boyutunda kontrastlanan nodüler lezyon ve her iki lakrimal bezde büyüme saptandı. Aksiller lenf nodu biyopsisinde granülomatöz inflamasyon görüldü. Toraks BT'de lenfadenopati, perilenfatik nodüller ve kemik tutulumunu düşündüren litik kaburga lezyonları izlendi. Yüksek ACE düzeyleri tanıyı destekledi. Klinik, radyolojik ve histopatolojik bulgulara dayanarak nörosarkoidoz ile birlikte multisistemik sarkoidoz tanısı konuldu. Bu olgu, özellikle sistemik tutulum varlığında, tek beyin lezyonu ve kraniyal nöropatiler olan hastalarda sarkoidozun ayırıcı tanıda düşünülmesi gerektiğini vurgulamaktadır.

**Anahtar Kelimeler**: Sarkoidoz, nörosarkoidoz, multisistem hastalık

## INTRODUCTION

Sarcoidosis is a multisystem granulomatous disease of unknown etiology that most commonly involves the lungs and intrathoracic lymph nodes.[1,2] Although extrapulmonary involvement is relatively common, central nervous system involvement, referred to as neurosarcoidosis, is rare and may present with diverse neurological manifestations, including cranial neuropathies and parenchymal brain lesions.

Radiological findings are often nonspecific and may mimic neoplastic or infectious conditions, particularly in cases presenting with solitary enhancing brain lesions. Therefore, the diagnosis requires integration of clinical, radiological, and histopathological findings.

Herein, we present a case of multisystem sarcoidosis with neurosarcoidosis presenting as facial nerve palsy and a solitary enhancing brain lesion.

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## CASE

A 41-year-old female presented with right-sided facial nerve palsy. Apart from right-sided facial nerve palsy, the neurological examination was otherwise unremarkable. No additional cranial nerve deficits, motor or sensory abnormalities, cerebellar signs, or meningeal signs were observed. Non-contrast cranial computed tomography demonstrated enlargement of the right lateral ventricle, prompting further evaluation with contrast-enhanced brain magnetic resonance imaging (MRI) and cerebrospinal fluid (CSF) flow study. MRI revealed asymmetric enlargement of the right lateral ventricle with leftward midline shift and periventricular edema suggestive of obstructive hydrocephalus. Irregular contrast enhancement was observed along the floor of the fourth ventricle at the level of the foramina of Magendie and Luschka, the pituitary stalk, and the floor of the third ventricle. Additional pial enhancement was noted at the level of the mesencephalon and pons. At the level of the foramen of Monro, enhancing lesions caused narrowing of the right foramen. Bilateral lacrimal gland enlargement with intense enhancement was also present. A 6 mm enhancing lesion was identified at the left frontoparietal junction (**Figure 1**). CSF flow through the aqueduct was preserved.

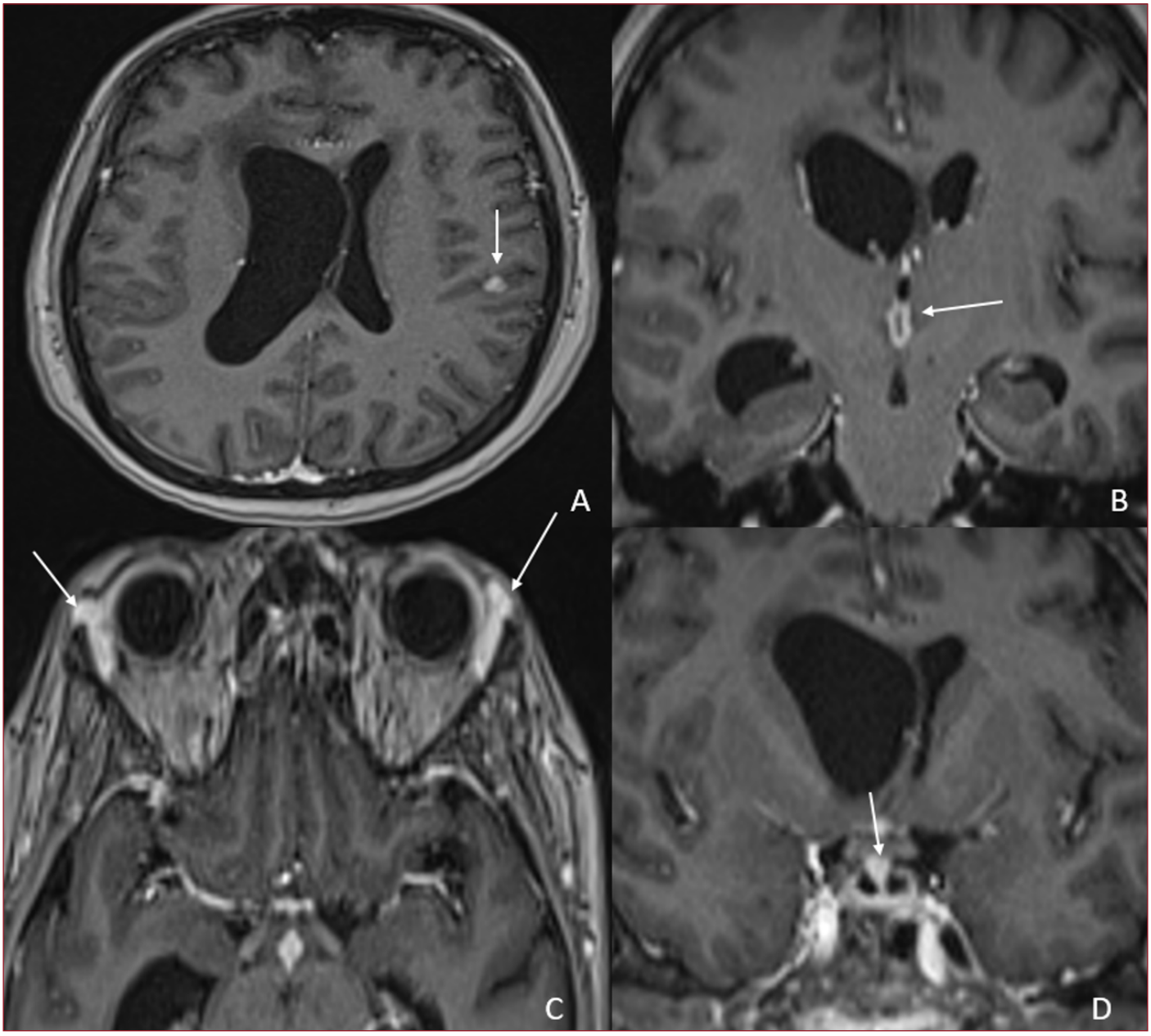


**Figure 1.** Contrast-enhanced brain magnetic resonance imaging (MRI). (A) Axial image demonstrates a small enhancing lesion in the left frontoparietal region (arrow). (B) Coronal image shows thickening and increased enhancement along the aqueduct of Sylvius and the wall of the third ventricle (arrow). (C) Axial image reveals bilateral lacrimal gland enlargement with intense enhancement (arrows). (D) Coronal image demonstrates thickening and enhancement of the pituitary stalk (arrow).

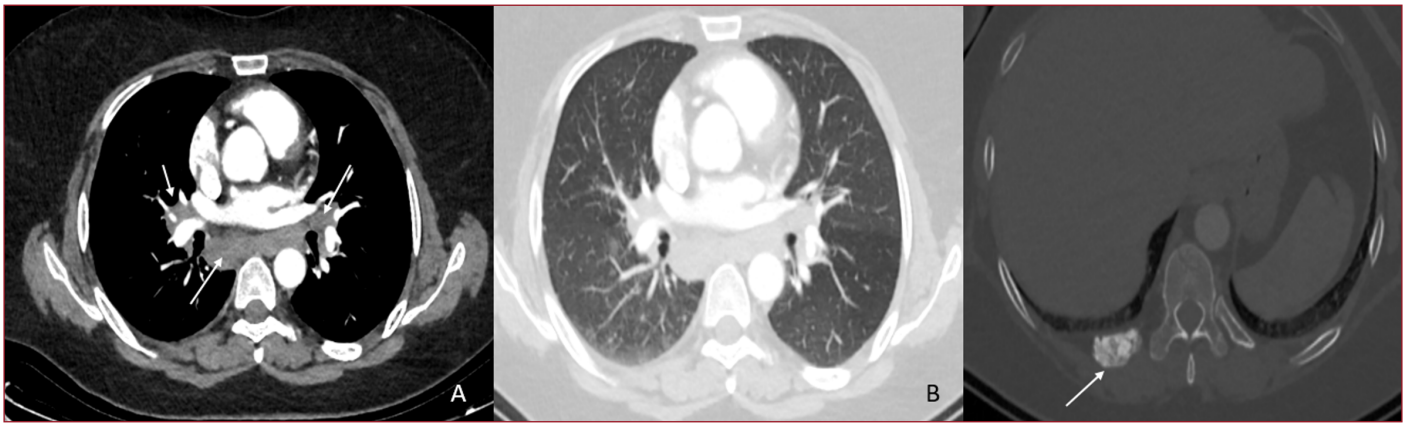


**Figure 2:** Contrast-enhanced thoracic CT findings in a patient with suspected neurosarcoidosis.(A) Axial contrast-enhanced CT image demonstrates multiple enlarged mediastinal and bilateral hilar lymph nodes (arrows).(B) Lung window image shows numerous perilymphatic nodules, predominantly in the right lung. (C) Bone window image reveals a sclerotic, expansile, and partially destructive lesion in the posterolateral arc of a rib (arrow).

Given these imaging findings, neurosarcoidosis was suspected, prompting contrast-enhanced thoracic computed tomography (CT). CT demonstrated mediastinal and hilar lymphadenopathy, along with perilymphatic nodules, more prominent in the right lung. In addition, a sclerotic, destructive, expansile lesion was identified in the posterolateral arc of the rib (**Figure 2**).

Elevated angiotensin-converting enzyme (ACE) levels (221 U/L) further supported the diagnosis. Extensive microbiological investigations were performed to exclude infectious etiologies. Acid-fast bacilli staining and mycobacterial PCR were negative. Fungal studies, including Aspergillus PCR and galactomannan antigen testing, were also negative. In addition, Pneumocystis jirovecii PCR and cytomegalovirus (CMV) PCR from bronchoalveolar lavage samples were negative. Concomitant axillary lymph node biopsy was performed under ultrasound guidance using a tru-cut technique due to a palpable axillary mass. Histopathological examination revealed necrotizing granulomatous lymphadenitis. Ziehl–Neelsen staining showed no acid-fast bacilli, and Periodic acid–Schiff (PAS) staining demonstrated no specific features.

Cerebrospinal fluid analysis was not performed in this case, which may be considered a limitation. However, the diagnosis was supported by characteristic clinical, radiological, and histopathological findings, along with the exclusion of infectious etiologies.

## DISCUSSION

The diagnosis of sarcoidosis relies on a combination of clinical, radiological, and histopathological findings, along with the exclusion of alternative granulomatous and infectious diseases.[1-4] Neurosarcoidosis is an uncommon but potentially severe manifestation, often presenting with heterogeneous imaging features that may mimic neoplastic or infectious conditions.[4-6] The multisystemic nature and variable clinical presentation of sarcoidosis, which may complicate the diagnostic process, have also been emphasized in the national literature.[7]

The differential diagnosis of enhancing central nervous system lesions in this setting is broad and includes neoplastic, inflammatory, and infectious etiologies. Primary central nervous system lymphoma and metastatic disease may present with enhancing parenchymal lesions and should be considered, particularly in patients with solitary or atypical brain lesions.[8]

Cranial nerve involvement is one of the most common manifestations of neurosarcoidosis, with facial nerve palsy being the most frequently affected cranial neuropathy. It may occur as an isolated finding or as part of more widespread central nervous system involvement. In addition, involvement of the hypothalamic–pituitary axis is well described and may present with pituitary stalk thickening and contrast enhancement, as observed in our case. Osseous involvement, although less common, has also been reported in sarcoidosis and may manifest as lytic or sclerotic lesions, particularly affecting the ribs and axial skeleton. The coexistence of cranial neuropathy, pituitary involvement, and bone lesions further supports the diagnosis of multisystem sarcoidosis and highlights the diverse clinical spectrum of the disease.[3,4,8-10]

However, in the present case, the presence of bilateral hilar and mediastinal lymphadenopathy, perilymphatic pulmonary nodules, lacrimal gland enlargement, pituitary stalk involvement, and histopathological evidence of granulomatous lymphadenitis supported a systemic granulomatous disease rather than a primary neoplastic process. Infectious granulomatous diseases, especially tuberculosis and fungal infections, are also important mimickers of neurosarcoidosis. Therefore, these entities were carefully evaluated and excluded by negative acid-fast bacilli staining, mycobacterial PCR, Aspergillus PCR, galactomannan antigen testing, Pneumocystis jirovecii PCR, and CMV PCR. Taken together, the clinical, radiological, histopathological, and microbiological findings favored probable neurosarcoidosis over lymphoma, metastatic disease, or infectious processes.

Central nervous system involvement may manifest with cranial neuropathies, parenchymal lesions, meningeal enhancement, or hypothalamic–pituitary axis involvement, as observed in the present case. In particular, facial nerve palsy is among the most common neurological manifestations and may be the initial presenting feature. Radiological findings such as leptomeningeal enhancement, ventricular involvement, and enhancing parenchymal nodules further contribute to diagnostic suspicion but are not pathognomonic.

In this case, the presence of characteristic thoracic findings, multisystem involvement, and histopathological evidence of granulomatous inflammation supported the diagnosis.

Although non-caseating granulomas are the hallmark of sarcoidosis, necrotizing granulomatous inflammation may rarely be encountered and should prompt careful evaluation for alternative diagnoses. In particular, infectious etiologies such as tuberculosis and fungal infections, as well as granulomatosis with polyangiitis, must be excluded. In the present case, microbiological and histochemical analyses, including Ziehl–Neelsen and PAS staining, were negative, and no clinical or laboratory findings supported an alternative diagnosis. Therefore, the diagnosis of sarcoidosis was established based on the overall clinical, radiological, and histopathological context. This case emphasizes the importance of considering sarcoidosis in the differential diagnosis of atypical central nervous system lesions, particularly when accompanied by systemic manifestations.

## CONCLUSION

Sarcoidosis should be considered in the differential diagnosis of atypical central nervous system lesions, as early recognition of its multisystem nature is crucial to avoid misdiagnosis and unnecessary interventions.

## ETHICAL DECLARATIONS

**Informed Consent**: Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

**Referee Evaluation Process**: Externally peer-reviewed.

**Conflict of Interest Statement**: The authors have no conflicts of interest to declare.

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**Author Contributions**: All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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