

# Erdheim-Chester Disease Presented with Bilateral Carotid Artery Occlusion: Case Report

## Bilateral Karotis Arter Oklüzyonu ile Prezente Olan Erdheim-Chester Hastalığı: Olgu Sunumu

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### ÖZET

Erdheim-Chester hastalığı etyolojisi bilinmeyen sistemik histiyositozisin nadir bir non-Langerhans formudur. Hastalık, muskuloskeletal sistem, kardiyak, pulmoner, gastrointestinal ve santral sinir sistemlerini de içeren multipl organ sistemini etkiler. Nörolojik belirtiler ise oldukça seyrekdir. Bu yazıda, bilateral kavernöz sinüs infiltrasyonu ve bilateral karotis oklüzyonu bulunan 44 yaşında bir Erdheim-Chester hastası sunulmuştur.

**Anahtar Kelimeler:** Erdheim-Chester hastalığı, oftalmopleji, karotis arterleri.

### ABSTRACT

#### Erdheim-Chester Disease Presented with Bilateral Carotid Artery Occlusion: Case Report

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Erdheim-Chester disease is a rare, non-Langerhans form of systemic histiocytosis of unknown etiology. The disease affects multiple organ systems, including musculoskeletal, cardiac, pulmonary, gastrointestinal, and central nervous systems, producing protean manifestations. Neurological manifestations are less frequent. We present a 44-year-old patient with Erdheim-Chester disease who had bilateral carotid artery occlusion and cavernous sinus infiltration.

**Key Words:** Erdheim-Chester disease, ophthalmoplegia, carotid arteries.

## INTRODUCTION

Erdheim-Chester disease is a rare, granulomatous and infiltrative disorder with proliferation of cholesterol-containing histiocytes and peculiar bone involvement (1). Erdheim-Chester disease is characterized by diffuse histiocytic infiltration of end organs with a clinical picture ranging from asymptomatic disease to a fulminant organ failure (2). The disease affects multiple organ systems, including musculoskeletal, cardiac, pulmonary, gastrointestinal, and central nervous systems, producing protean manifestations (3-6). Neurological manifestations are less frequent, and the most frequent central nervous system manifestations are diabetes insipidus, cerebellar syndromes, orbital lesions, and extra-axial masses involving the dura (3,7). In this paper, we present a 44-year-old patient with Erdheim-Chester disease who had bilateral carotid artery occlusion and cavernous sinus infiltration.

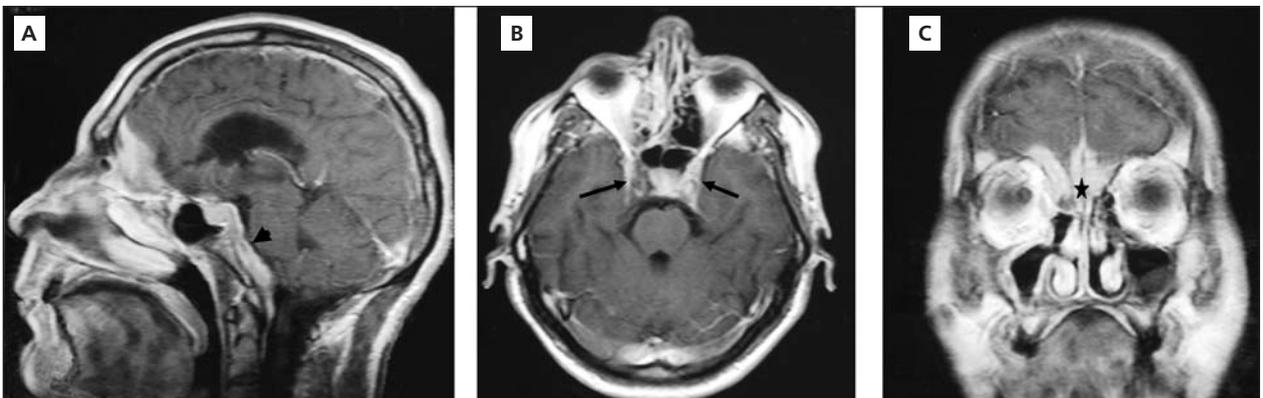
## CASE

A 44-year-old male presented with double vision, difficulty in walking and paresthetic complaints on his arms and legs bilaterally. He had double vision for one year, and this symptom was followed by difficulty in walking and sensorial complaints for the last two months. On neurological examination, he was awake and alert, and his speech was fluent. Pupils were unequal, with the right pupil larger than the left; both pupils were reactive to light and accommodation. Fundoscopic examination was normal. Bilateral horizontal eye movements were limited and vertical eye movement was limited on the

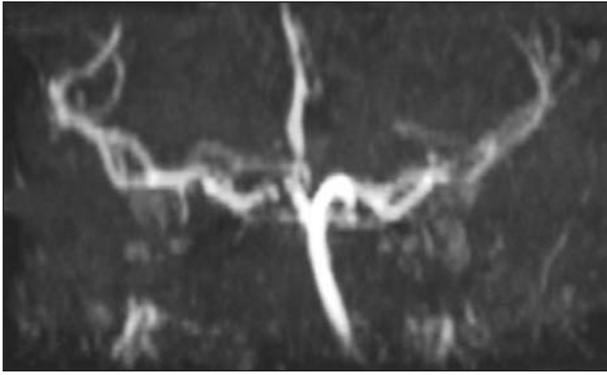
right side. The examination of other cranial nerves was normal. Although muscle strength of all muscles was 5/5, tendon reflexes were 3+/4, and plantar reflexes were extensor bilaterally. Sensation was intact throughout the tests of light touch, pinprick, vibration, proprioception, and temperature. He was able to walk on his heels and toes.

Routine blood chemistry, blood angiotensin converting enzyme (ACE) level and blood antibody titers were normal. Sagittal, axial and coronal T1-weighted cranial magnetic resonance (MR) images showed thickening of epidural soft tissue in the anterior cranial fossa and prepontine cistern as well as infiltration of bilateral cavernous sinuses (Figure 1). Cranial MR angiography (MRA) showed signal loss of bilateral internal carotid artery due to the infiltration of the cavernous sinus (Figure 2). Frontal craniectomy was performed and immunohistochemical examination of the epidural region revealed positive CD68 and negative CD1a and S-100, and these results were specific for Erdheim-Chester disease (Figure 3).

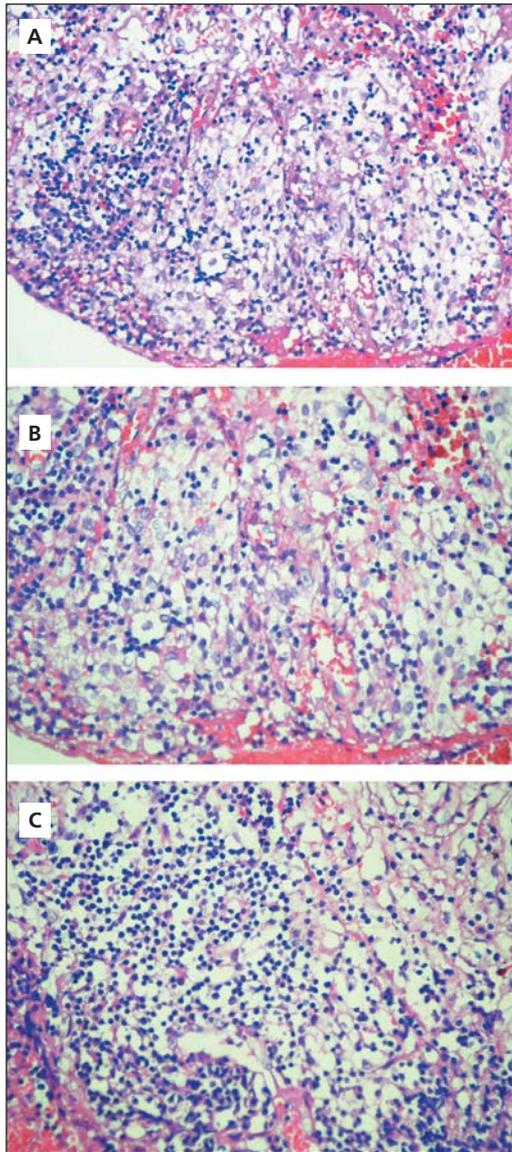
Methylprednisolone (60 mg per day) treatment was given, and four months later clinical findings had partially recovered, and horizontal and vertical gaze movements were improved. Muscle strength of all muscles was nearly normal. His walking difficulty had also improved four months later. Two years later, follow-up MR was nearly normal but MRA showed bilateral carotid artery occlusion, and he had spastic gait.



**Figure 1.** Sagittal contrast-enhanced T1-weighted images show epidural soft tissue thickening on the anterior cranial fossa (star) and pre-pontine area (arrowhead). Also note the presence of bilateral cavernous sinus infiltration.



**Figure 2.** Magnetic resonance angiography reveals no signal from bilateral internal carotid arteries due to bilateral cavernous infiltration.



**Figure 3.** Immunohistochemical slices of the epidural region revealing histiocytic cell infiltration.

## DISCUSSION

Erdheim-Chester disease is a rare non-Langerhans cell histiocytosis (1-3). The disease is characterized by tissue infiltration by lipid-laden macrophages, multinucleated giant cells, and inflammatory cells composed of lymphocytes and histiocytes, typically in the bone marrow, but also in numerous other organs (8).

Neurological disorders due to Erdheim-Chester disease occur in the course of the illness and are most often associated with intracranial tissue infiltration by foamy histiocytes (9-12). The lesions of the central nervous system occur most often in the cerebellum and pons, and involvement of the cerebral hemispheres, pituitary stalk, hypothalamus, brain stem, choroid plexus, spinal dura and falx may also be seen (11,12). The most common neurological presentations of Erdheim-Chester disease are diabetes insipidus, cerebellar signs, paraparesis, seizures, and orbital involvement (12). Ophthalmological findings in Erdheim-Chester disease are xanthelasma, thinning of lids, exophthalmos, ophthalmoplegia, optic disc swelling, optic atrophy, and retinal striae (9). Our case presented with ophthalmoplegia due to cavernous sinus involvement.

It is known that Erdheim-Chester disease may involve the cardiovascular system (13). Haroche et al. analyzed 72 patients with Erdheim-Chester disease who had cardiovascular involvement: 40 (55.6%) of them had periaortic fibrosis, 32 (44.4%) had pericardial involvement, and 22 (30.6%) had myocardial involvement (13). Gauvrit et al. illustrated a rare case of pericarotid fibrosis in a patient with Erdheim-Chester disease (14). In our patient with ophthalmoplegia, we demonstrated Erdheim-Chester disease with pathological findings, and we showed bilateral carotid artery occlusion due to pericavernous infiltration on MR imaging and MRA.

In conclusion, in patients with ophthalmoplegia, especially in young patients with other neurological, neuroradiological and systemic findings, Erdheim-Chester disease should be taken in consideration as a rare clinical manifestation.

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