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Case Report

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Osteopetrosis of temporal bone causing bilateral peripheral facial nerve paralysis: MRI and CT findings

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Abstract

Osteopetrosis is a rare, heterogeneous group of genetic disorders characterized by osteoclast dysfunction, resulting in excessive bone sclerosis. When it affects the skull base and temporal bone, it can lead to Eustachian tube dysfunction, recurrent otitis media, hearing impairment, and cranial nerve deficits. This report highlights the computed tomography (CT) and magnetic resonance imaging (MRI) findings of temporal bone osteopetrosis in an 18-year-old male presenting with acute-onset bilateral facial nerve paralysis. Radiological evaluation revealed significant narrowing of the bilateral internal acoustic canals, a hallmark feature of the disease. The patient received a short course of corticosteroid therapy, resulting in complete resolution of symptoms. This case underscores the critical role of imaging in diagnosing and managing cranial nerve complications in temporal bone osteopetrosis.

Keywords: computed tomography; magnetic resonance imaging; temporal bone; osteopetrosis; facial paralysis

1. Introduction

Osteopetrosis is a rare genetic bone disorder characterized by sclerosis of the bones resulting from defective osteoclast function. This dysfunction impairs the resorption of bone and mineralized cartilage, leading to dense but fragile bones that are prone to fractures and other complications (1). The involvement of the skull base and temporal bone in osteopetrosis can result in a variety of clinical manifestations, including Eustachian tube dysfunction, recurrent otitis media, hearing loss, and cranial nerve deficits (2,3). Such cases require thorough radiological assessment to guide diagnosis and management. Herein, we report a case of osteopetrosis in an 18-year-old male presenting with acute-onset bilateral facial nerve paralysis, highlighting the computed tomography (CT) and magnetic resonance imaging (MRI) findings associated with this rare presentation.

2. Case presentation

An 18-year-old male patient with a known history of osteopetrosis presented to hospital with complaints of acute-onset bilateral facial nerve paralysis. Laboratory investigations, including hemoglobin levels and white blood cell counts, were within normal limits. On physical examination, there was notable weakness of the facial muscles on both sides, affecting the forehead and lower face symmetrically. Otoscopic examination revealed intact tympanic membranes and normal external auditory canals, with no evidence of hearing loss. High resolution CT of the temporal bones showed diffuse sclerosis involving the temporal bones and skull base (Fig. 1). Notably, there was significant narrowing of the bilateral internal acoustic canals. MRI of the same region demonstrated decreased signal intensity of the affected bones on both T1-weighted (T1W) and T2-weighted (T2W) sequences, consistent with sclerotic changes (Fig. 2). The patient was admitted to the hospital and initiated on a three-day course of corticosteroid therapy. His symptoms resolved entirely during hospitalization, and he was discharged in stable condition with no residual facial nerve deficits.

3. Discussion

Osteopetrosis encompasses a heterogeneous group of heritable bone disorders characterized by impaired bone resorption due to osteoclast dysfunction leading to bone sclerosis (1). This dysfunction may result from defective differentiation of hematopoietic stem cells into osteoclasts or from a failure in the function of already differentiated osteoclasts. Although the affected bones appear radiologically dense, they are structurely fragile and prone to fractures, highlighting the paradoxical nature of this disorder (4).

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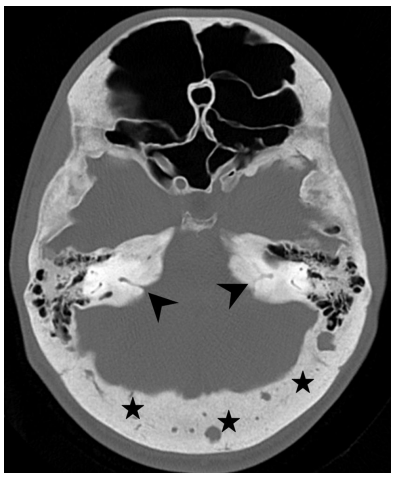

Fig. 1. Axial CT image of the temporal bone showing diffuse sclerosis involving the temporal bones and entire skull base (asterix). Note the significant narrowing of both internal acoustic canals (arrowheads).

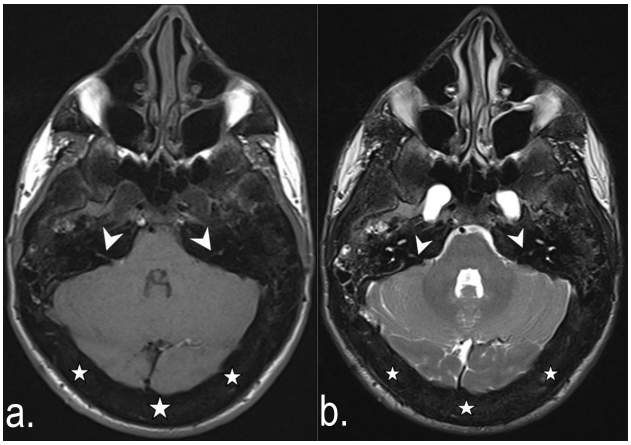

Fig. 2. T1W and T2W images of the temporal region reveal pronounced signal loss in the medullary bone (asterisk), indicative of extensive sclerosis. Severe narrowing of the bilateral internal acoustic canals is also evident (arrowheads), correlating with the radiological findings on CT.

The clinical presentation of osteopetrosis varies based on its genetic subtype. Autosomal-dominant osteopetrosis, commonly diagnosed in young adults, generally has a more favorable prognosis and is divided into type I and type II. Type I typically spares the skull base and temporal bones, while type II frequently involves these regions (3). By contrast, the autosomal-recessive form, also known as malignant osteopetrosis, has a more severe course and manifests in infancy with higher morbidity and mortality.

Temporal bone involvement in osteopetrosis can lead to significant clinical complications, including stenosis of both external and internal auditory canals, ossicular ankylosis, vascular narrowing, Eustachian tube dysfunction, otitis media, sensorineural hearing loss, and cranial nerve deficits (2,3). Facial nerve compression, as seen in this case, may manifest as peripheral facial nerve paralysis, clinically resembling Bell's palsy.

Imaging plays a pivotal role in diagnosing temporal bone osteopetrosis. High-resolution CT typically reveals diffuse sclerosis, obliteration of medullary cavities, reduced diploic spaces, and narrowing of auditory canals (3). The stapes may also appear thickened in some cases. On MRI, sclerotic bone is characterized by decreased signal intensity on both T1W and T2W sequences.

Currently, there is no definitive preventive treatment for osteopetrosis. Management is primarily symptomatic and supportive. Cranial nerve dysfunction, including facial nerve paralysis, may be addressed with decompression surgery, although evidence supporting its efficacy remains limited (4,5). In cases of severe sensorineural hearing loss, cochlear implantation can provide functional improvement (6).

In conclusion, temporal bone osteopetrosis is a rare but clinically significant manifestation of this complex disorder. The disease frequently presents with increased bone density and extensive sclerosis, leading to complications such as hearing loss and facial paralysis. This case highlights the characteristic CT and MRI findings in an 18-year-old male with osteopetrosis.

Conflict of interest

The authors declare that they have no competing interests.

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Informed consent was obtained for this case presentation.

Authors' contributions

Concept: M.Ö., A.T.S., Ç.Ç., Design: M.Ö., A.T.S., Data Collection or Processing: M.Ö., A.T.S., Analysis or Interpretation: M.Ö., A.T.S., Literature Search: M.Ö., A.T.S., Writing: M.Ö., A.T.S., Ç.Ç.

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