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Hypertelorism

Mucocele of the paranasal sinuses as a cause of acquired orbital hypertelorism: the second case

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Abstract

Background: Orbital hypertelorism is defined as a lateralization of the total orbit. This condition is associated almost always with congenital abnormalities. An acquired total displacement of the orbit is almost impossible once the bony pillars of the craniofacial complex are established. The aim of this article was to discuss a case of a young man with paranasal sinus mucocele, who developed OHT.

Case Description: A 20-year-old man was admitted to us with orbital hypertelorism in 1993. The patient's examination revealed marked orbital hypertelorism, and no other abnormal finding was noted, and he had no congenital defect at birth. Hypertelorism was corrected by surgery, and we find that this condition was acquired secondary to paranasal sinus mucocele.

Conclusion: This is the second case of an acquired OHT secondary to paranasal sinus mucocele and the first in which the condition was corrected by surgery in late years of life. We found excellent correction of orbital hypertelorism 13 years after surgery. The authors discuss the possible link between mucocele and possible mechanism for the development of orbital hypertelorism and emphasize the need for early surgery and long-term follow-up.

Keywords: Hypertelorism; Mucocele; Paranasal sinus

1. Introduction

The term *ocular hypertelorism* was first introduced by Greig [6] as "a widening of the interpupillary distance," but then, it had been widely accepted that the bony distance between the medial bony walls of the orbis or the distance between the inner canthi to be one of the most reliable measurement for defining the OHT [4,10]. Series including normal population showed that this distance normally averages 16 ± 4 mm at birth, 25 ± 4 mm at 12 years of age, and 35 ± 4 mm in adults [3]. OHT was classified into 3 degrees according to the roentgenographic measurements of interorbital distance by Tessier [15], and it is the third degree that indicates the interorbital distance exceeding 40 mm, which includes gross facial disfigurements for which surgical intervention is indicated.

It is well known that hypertelorism seldom exists by itself and is mostly associated with congenital defects caused by the midline closure defects along the neuroaxis and various degrees of intracranial developmental anomalies [4,8-10]. Encephaloceles deforming the malar, sphenoidal, and frontal bones; brachycephaly stretching the supraorbital arch and flattening the supraorbital rims; gigantic frontal bone pneumatization; and widened ethmoidal cells displacing the orbis laterally are the most common anomalies associated with OHT.

The English literature provided us with only 1 case of an adolescent with mucocele of the paranasal sinuses as a cause of acquired OHT, presented by Jackson et al [8]. This is the second case of an adolescent who developed OHT due to mucoceles of the paranasal sinuses. We emphasize

Abbreviations: CT, computed tomography; ENT, ear-nose-throat; MRI, magnetic resonance imaging; OHT, ocular hypertelorism; URTI, upper respiratory tract infections.

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Fig. 1. This photography was taken in 1974 when he was 1 year old. There was no abnormal appearance on his face or head (arrow). We publish this photograph with the permission of the patient and his family.

the association between mucocele and development of acquired OHT. Furthermore, we want to underline the need for early surgical correction and the need for the long-term follow-up.

2. Case report

In 1993, a 20-year-old man was presented for investigation of progressive OHT. His parents stated that the patient had no congenital defect at birth and had no abnormal finding during his early childhood (Fig. 1). His growth and development were initially normal, and general health had been well until the age of 8 years. The remarkable finding from the history was that the patient developed multiple URTI and headaches. In spite of treatment, several hospital admissions were required because of URTI caused by influenza virus. His facial features had chanced progressively, and nasal obstruction with mucoid discharge began when he was 8 years old (Fig. 2). At that time, the patient was operated in an ENT clinic twice, and the physicians said to his family that the patient would die within 2 months. We have no information related to histopathological diagnosis of the specimens extracted at that time. For 12 years, the patient's head and facial features changed remarkably and included marked OHT associated with monstrous deformity of the face, in addition to the severe eye dysfunction.

On presentation, the physical examination revealed a flattened nasal bridge with marked OHT (Fig. 3A and B). Intercanthal distance was 48 mm; interpupillary distance, 75 mm; and interlateral canthal distance, 90 mm. The level of the palpebral fissures was not on the same plane. In addition, there was marked nasal deviation to the left. Intranasal examination showed bilateral nasal obstruction and pale edematous mucosa with a mucoid discharge. MRI showed extensive mucocele involvement of ethmoid, frontal, and sphenoid sinuses (Fig. 4a). A CT reconstruction imaging clearly demonstrated the abnormal bony shapes caused by mucocele (Fig. 4b). The remainder of the physical examination was negative.

The patient underwent operation in which intracranial approach for correction was required. A bicoronal (ear-to-ear) scalp flap was elevated and turned forward over the face. A frontal bone flap was performed; a supraorbital bar was maintained bilaterally. After the removal of the frontal

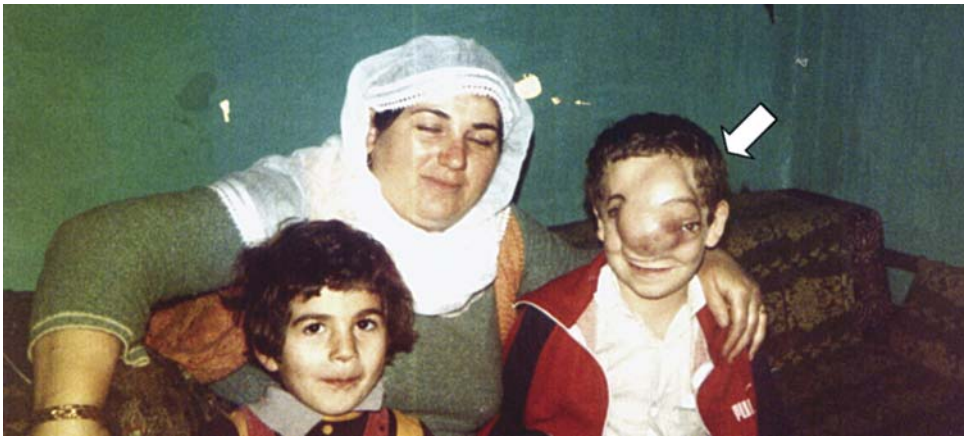

Fig. 2. The patient when he was 8 years old before ENT surgery. Note marked frontonasal mucocele, exotropia, and hypertelorism. This photograph is published with permission from the patient and his family.