

Unilateral Congenital Third Nerve Palsy Associated with Osteogenesis Imperfecta

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Abstract

Purpose: Congenital third nerve palsy was generally thought to exist in isolation without associated abnormalities. In this report, we present a case of congenital third nerve palsy in osteogenesis imperfecta.

Methods: A 6-month-old girl with osteogenesis imperfecta presented with ptosis and large incomitant exotropia in left eye (LE) at birth. Ocular examination revealed total third nerve palsy in LE. Brain CT scan was normal. She underwent surgery for strabismus at the age of two years and surgery for ptosis after four months.

Results: Ocular alignment has greatly improved after recess-resect procedure on the horizontal rectus muscles in LE. She achieved a good functional outcome after ptosis surgery.

Conclusion: Congenital third nerve palsy may associate with systemic disease. To our knowledge, this represents the first report on demonstrating this association in osteogenesis imperfecta.

Keywords: Congenital Third Nerve Palsy, Osteogenesis Imperfecta

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Introduction

Unlike acquired third nerve palsy in childhood, the congenital form is usually considered to be an isolated disorder, not associated with other neurologic or systemic disorders.¹

Osteogenesis imperfecta (OI) is an inherited connective tissue disorder that primarily affects the bone. OI is a genetic disorder resulting from an abnormal quantity and/or quality of type I collagen and is classified into four type.²

The ocular complication in this disease consist of blue sclera, fragile cornea, retinal hemorrhage and spontaneous scleral rupture.³⁻⁶ Here we present a case of congenital third nerve palsy in OI.

Case report

A 6-month-old girl with OI was referred to strabismus service at Nikookary Eye Hospital in Tabriz due to ocular deviation and ptosis in left eye (LE) at birth. The patient, born at 37 weeks gestation by normal delivery, was diagnosed with OI at the age of one month due to multiple fractures in radius and tibia and radiologic manifestations (Figure 1). The family history is negative. Ocular examination revealed severe ptosis, large exotropia, no adduction, no elevation no depression, miotic pupil with no reaction to light, and blue sclera in LE. The diagnosis was congenital third nerve palsy in LE (Figure 2).

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Figure 1. Leg deformity due to multiple fractures



Figure 2. Total third nerve palsy in left eye

Neurologic examination and brain CT scan was normal. Her first surgery was performed at the age of 24 months due to the risk of odontoid process fracture during intubation in general anesthesia (10 mm recession of lateral rectus and 7 mm resection of medial

rectus in LE). Postoperatively orthotropia was achieved in primary position.

This procedure facilitated treatment of amblyopia. Because of occlusion of visual axis, frontalis suspension with silicon rod was performed after four months.

She achieved a good functional outcome after this procedure (Figure 3).



Figure 3. After strabismus and ptosis surgery

Discussion

Third cranial nerve palsy in children is uncommon and often congenital (caused by adverse intrauterine events, a traumatic delivery, neonatal hypoxia or the result of postnatal trauma, neoplasia, migraine and infection).^{2,7,8}

The most frequently cited mechanism in congenital form is perinatal injury to the peripheral third nerve.⁹ The majority of these children are otherwise healthy.¹⁰ Although the congenital form is usually considered to be an isolated disorder, not associated with other neurologic or systemic disease, but various neurological abnormalities can accompany congenital third nerve palsy.^{1,10} These consist of congenital absence of the nerve and/or nucleus, midbrain or cerebellar infarcts, midline CNS defects, cerebral ventricular abnormalities, hypoplasia of the midbrain and corpus callosum.^{9,10,11}

The other neurologic abnormality associated with palsy is septo-optic dysplasia.^{9,11} Therefore all children with third nerve palsy require neuroimaging. In our case, brain CT scan and neurologic examination was normal, although she was a known case of OI.

OI is the most common inherited connective tissue disorder that primarily affects bone.

However, it is a systemic disorder, evidenced by the occurrence of ocular complication, dentinogenesis imperfecta, hearing loss, joint laxity, restrictive pulmonary disease, and short stature.³

In our case, the patient had third nerve palsy in LE. Patients with OI may have an accompanying bleeding tendency secondary to capillary fragility caused by abnormal collagen support around blood vessels.² These factors may predispose the patient to peripheral nerve injury in the setting of minor trauma in perinatal or at birth.

Pupil in congenital third nerve palsy may be spared or may be involved (dilated or miotic). Pupillary miosis is a result of aberrant reinnervation and appears to be much more frequent after congenital than acquired palsy.¹

In our case pupil in LE was miotic with no reaction to light.

Conclusion

Here we present a case of congenital third nerve palsy with OI that to our knowledge has not been reported previously. Minor trauma in perinatal or in delivery may be the etiologic mechanism for nerve injury in OI.

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