CASE REPORT

Congenital Facial Paralysis: Facial Nerve Hypoplasia

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Introduction

Facial paralysis in a newborn is called congenital facial paralysis (CFP). Two most common etiologies of CFP are birth trauma and developmental [1]. Differentiation between birth trauma and developmental causes of CFP is the most important issue in planning the management and informing the parents about the prognosis. This can be achieved by history and physical examination in some cases, however diagnosis may remain uncertain despite all efforts. We herein report a case of CFP, having a normal computed tomography (CT) of the temporal bone whereas a severely hypoplasic facial nerve was seen on the magnetic resonance imaging (MRI) which helps to determine the congenital etiology.

Case Report

A 7-months-old boy was presented with left-sided facial paralysis noticed at birth. There was no history of birth trauma or facial paralysis in his family. He had facial paralysis affecting all divisions of facial nerve, however, function at the oral commissure improved partially afterwards. His face was symmetric at rest. He also had right clubfoot. Rest of the otolaryngologic and systemic examinations was normal. He was found to have normal hearing bilaterally with transient evoked otoacoustic emissions, which was confirmed later with pure tone audiometry. EMG revealed severe axonal lesion on the left side, left orbicularis oris muscle was found to be innervated by the right facial nerve. Temporal bone and facial canal were found to be normal in the computed tomography (CT) (Figure 1). However magnetic resonance imaging (MRI) scans revealed a severely hypoplastic facial nerve at cysternal and canballcular segments (Figure 2). No improvement in the facial functions was observed in 2 years of follow-up.
Discussion

Congenital facial paralysis is an uncommon situation with an occurrence of 2 in 1,000 alive births. The major role of the physician is to differentiate the etiology of the facial paralysis whether is traumatic or developmental. This distinction is important not only for treatment options and prognosis but may have medicolegal implications as well [2]. Traumatic CFP may be diagnosed by the history and physical examination. Complicated delivery, prematurity, ecchymosis around ear, facial paralysis affecting mostly lower division are factors associated with traumatic CFP. Recovery even with synkinesis is the rule in traumatic cases.

Developmental CFP may be diagnosed more easily when it is part of a syndrome. CFP may be a related to a number of syndromes such as Mobius syndrome, velocardiofacial syndrome, Goldenhar’s syndrome and Poland’s syndrome [3]. Otologic, craniofacial and systemic anomalies should be evaluated carefully in a patient born with facial paralysis. Bilateral involvement, other cranial neuropathies also imply developmental etiology. Predominance of upper or lower divisions may be seen. Facial tonus is usually good in these cases. However, when the CFP occurs as an isolated incident the diagnosis may be more challenging.

In our case isolated unilateral CFP in the child of a primiparous mother supports a traumatic cause of CFP. On the other hand there was no finding such as ecchymosis, hemotympanum in physical examination. No recovery in facial nerve functions emphasizes developmental etiology. Late admission in this case lessens the value of the electrophysiologic tests. Although there were controversial findings in the EMGs, no sign of reinnervation is thought to be compatible with developmental origin. Contralateral innervation of the facial nerve may be seen in normal people but, its incidence increases significantly after facial nerve injury [4].

Absence of facial canal may be recorded in CT scan despite a visible facial nerve in internal auditory canal (IAC) [5]. On the other hand facial nerve may be abnormal in spite of a normal CT as seen in the present case. Brainstem hypoplasia, abnormal facial nucleus, aplasia of the facial nerve may be seen in MRI with a normal or abnormal CT [6,7]. Internal auditory canal absence or stenosis may also be revealed in CT or MRI. Even in those cases, facial nerve function is intact in the majority of the cases, whereas vestibulocochlear remains dysfunctional [8].

The structure which is seen as severely hypoplastic facial nerve in the MRI may represent the fibrous cord found in the surgical explorations of the CFP [9].

Traumatic CFP has 90% chance of spontaneous recovery. Although being controversial, steroid treatment may be considered in a case diagnosed with traumatic CFP [10]. Surgical exploration may also be
indicated in cases with rapid deterioration. This good prognosis is not shared by developmental cases. Surgical exploration is discouraged in developmental cases because the facial nerve is absent or like a fibrous cord. Nerve transpositions are useless in CFP cases leaving only regional or microneurovascular muscle transfers and static techniques as the treatment alternatives.

This case shows the MRI examination has a crucial role in the evaluation of patients with CFP and should be present in the diagnostic battery of the CFP. CT findings may be normal whereas, a hypoplastic facial nerve on MRI may help us to attain definite diagnosis and direct the clinical management and inform the parents about the prognosis of the disease.

References