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An unusual cause of facial nerve palsy affecting a mum and her child with literature review and analysis for other causes

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ABSTRACT

Facial nerve palsy is a condition with several implications, particularly when occurring in infancy and childhood. It represents a clinical problem with regard to its etiology, treatment options and its outcome, particularly when occurs in infants and children. There are several causes of facial nerve paralysis in children, as it can be congenital (due to delivery traumas and genetic or malformation diseases) or acquired (due to infective, inflammatory, neoplastic, traumatic or iatrogenic causes). Nonetheless, 40-75% of the cases of unilateral facial paralysis remain idiopathic.

Unilateral familial congenital facial nerve palsy is an extremely rare condition that is usually syndromic, namely, in Moebius syndrome. The occurrence of isolated familial facial nerve palsy is even rarer, with only a few cases reported in the literature. Here we report a mother and her child both have congenital facial paralysis on the same side.

Keywords: Facial paralysis, Seventh cranial nerve, Children, Bell's palsy,

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Introduction

Congenital facial paralysis [CFP] is uncommon and may cause multiple problems for the newborn, such as difficulty with nursing and incomplete eye closure leading to less eye protection. It is classified as unilateral or bilateral and complete or incomplete.

It Is important to differentiate between birth trauma-related and developmental causes of CFP because it serves as a cornerstone in planning the treatment and informing the parents about the prognosis. History and physical examination can guide the diagnosis.

There are many possible causes of facial nerve paralysis in children. These can be classified as congenital related to birth trauma, syndromic and non-syndromic malformations and genetic. Or acquired caused by infectious, inflammatory, neoplastic, traumatic conditions [1,2,3], [Table 1]. However, significant numbers of Pediatrics facial nerve palsy remain unknown. Bell's palsy is the most frequent form of facial paralysis both in adults and children [4].

An accurate diagnosis is necessary to assess the prognosis and the therapeutic options.

Table 1: Possible causes of facial nerve palsy in children

Idiopathic	Bell's palsy			
Congenital	Delivery traumas	Syndromic malformation	Genetic	
	Maternal primiparity, Birth weight greater than 3.5kg, Use of forceps, Cesarean birth and Prematurity.	Mobius syndrome Goldenhar syndrome Syringobulbia Arnold-Chiari syndrome	Hereditary myopathies	
Acquired	Infectious	Inflammatory	Neoplastic	Traumatic
	Ramsay Hunt syndrome, Epstein Barr virus, Haemophilus Influenza, Tuberculosis, Lyme disease, Cytomegalo and Adenovirus, Rubella, Mumps, Human immunodeficiency virus, Acute/Chronic Otitis media	Henoch-Schonlein purpura Kawasaki syndrome	Schwannoma of VII cranial nerve Hemangiomas Rhabdomyosarcoma Temporal bone histiocytosis Leukaemia Parotid gland tumours	Temporal bone fracture Iatrogenic

Clinical presentation

A 2-years old boy, Fully term-born, by normal vaginal delivery not involving forceps, was presented with a right side lower motor facial paralysis. In his parents' family, his mother 35-years old, was born with a similar right-sided congenital facial paralysis as well affecting all sectors of the right facial nerve. Physical examination for both showed a facial asymmetry, incomplete right eye closing, left deviation of the angle of the mouth, and right-sided loss of nasolabial furrow and Right forehead wrinkle. [photo 1, 2]

No impairment of other cranial nerve functions is founded in the boy and her mother, namely, there are no hearing, gait or mobility disorders on both [Cochlea-vestibular nerves are normal]. Brain MRI was normal, facial nerve conduction study showed severely reduce amplitude in the right facial nerve.

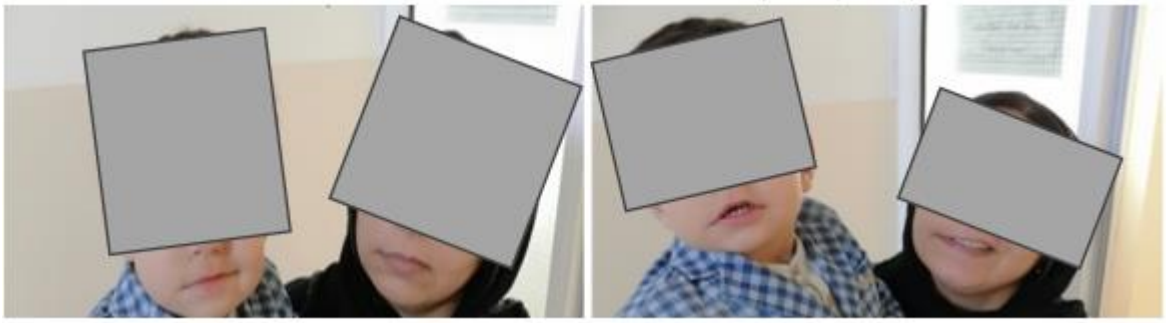

Figure 1&2: The boy and his mother show clear right lower motor facial nerve palsy

Diagnosis:

A comprehensive history is always important for the correct diagnosis. It is necessary to inquire about the onset and the time course of the paralysis and its eventual progression. For example a gradual onset for > 3 weeks may suggest a possibility of a neoplastic etiology. All the associated symptoms should be identified, as well as any other comorbidities affecting the child [1-5].

During the ENT examination, particular attention should be given to the inspection of the external auditory canal, the eardrum and the mastoid region. The facial nerve evaluation, in terms of facial movements and spontaneous expressions, should be classified according to the House-Brackmann grading system. Both the eye and the palpebral region as well as the lower face should be carefully observed at rest and movement with good documentation of the asymmetry [5,6]. The audiological evaluation is important to assess the presence of stapedial reflexes [topo-diagnosis] and eventually to document the presence of hearing loss [7].

Blood pressure and full blood count should be verified in all cases of pediatric paralysis. Particularly in children, it has been described that high blood pressure levels can be associated with recurrent facial palsy. Furthermore, a moderate increase of monocytes and lymphocytes has been found with Bell's palsy [8]. The lumbar puncture is performed when suspecting meningitis which is usually associated with severe headache, fever, papilledema and neck stiffness. While in Guillain-Barré syndrome, the analysis of the cerebrospinal fluid usually shows a characteristic increase in protein not accompanied by consensual cells increasing [albumin-cytological dissociation] [4,9].

Specific laboratory and imaging tests are not routinely indicated, but may help in patients with recurrent paralysis or when there has been no improvement after three weeks of therapy. For the Ramsey Hunt syndrome in children, an ELISA test for IgM and IgG antibody titer against Herpes Varicella-Zoster is recommended [8,10]. Serologic tests for Lyme disease should be considered in the endemic areas or for people travelling to these forest areas. while In case of clinical suspicion of a neoplastic aetiology, the computed tomography of the petrous bone and the brainstem magnetic resonance imaging must be performed. Radiological images are required when the child shows other neurological manifestations or in chronic otitis media, acute mastoiditis or temporal bone fracture [11-14].

Electrophysiological studies can be useful to identify the cause of the paralysis and to define the prognosis and follow-up of functional recovery, but they are still not considered necessary in all pediatric patients [1-6].

Discussion

There are many possible causes of facial nerve paralysis in children. These can be classified as congenital [traumatic, syndromic and non-syndromic malformations, genetic] or acquired [infectious, inflammatory, neoplastic, traumatic].

Unfortunately, in about 50% of the cases, the aetiology remains unknown: these forms are classified as Bell's palsy. In children, Bell's palsy has an estimated incidence of about 6.1 cases per year per 100,000 in those aged between 1 and 15 years [2-5]. It is believed that it can be caused by viruses such as Herpes simplex. About 70% of Bell's palsy has a favourable prognosis with spontaneous and complete resolution within 3 months. The paralysis severity at the onset can influence the degree of recovery; severe paralysis hardly obtains a complete recovery of nerve function [8].

Congenital facial paralysis can result from developmental defects or delivery traumas. Perinatal traumas are the most frequent cause of congenital paralysis. The main reported risk factors associated with traumatic facial paralysis are: the mother's first child, birth weight greater than 3500 g, use of forceps, cesarean birth and prematurity [4,5]. These cases have usually a favourable prognosis, with infants recovering the full functionality of the seventh cranial nerve within a few months without sequelae.

An isolated congenital facial nerve paralysis can be caused by the Möbius syndrome, as in our case here. In Möbius syndrome other cranial nerves such as the III, IV, V, and VIII can be involved, which is not the case in our patient. The reported prevalence of this syndrome is about 1/150,000 live births [1,2,4,8]. It is reported to be due to hypoplasia of the motor nuclei of the cranial nerve/s within the brainstem, probably due to hypoxic-ischemic encephalopathy.

Those affected by Goldenhar syndrome; hemifacial microsomia, with a spectrum of congenital malformations involving the structures derived from the first and second branchial arch, can also present a congenital facial paralysis [15].

Congenital pseudobulbar palsy [Syringobulbia] is a condition that clinically manifests with facial paralysis, dysphagia and speech difficulties. While in the Arnold-Chiari syndrome, congenital facial paralysis is usually associated with other cranial nerves paralysis; especially the fifth cranial nerve and it is due to malformations of the posterior fossa that allow herniation of brain structures through the foramen magnum. [16]

Genetic causes of facial nerve paralysis include hereditary myopathies, such as myotonic dystrophy and myasthenia [4,8].

Acquired facial paralysis can frequently be due to viral infections. The reactivation of Herpes Varicella-Zoster may be responsible, even in children, of Ramsay Hunt syndrome [Herpes Zoster Oticus] [10]. In such cases, facial palsy can be associated with the presence of vesicular lesions of the external auditory canal and/or of the auricular concha. The incidence of this syndrome under 10 years of age is reported to be 2.7/100,000. Not frequently, bilateral facial nerve palsy may be caused by Epstein-Barr virus infection, Haemophilus influenza, tuberculosis or Borrelia burgdorferi infection [17]. Other agents that may cause facial nerve palsy in children are cytomegalovirus, adenovirus, rubella, mumps, Mycoplasma pneumonia and HIV[1,4].

Facial nerve palsy may also be present as a complication of several diseases such as acute and chronic otitis media, cholesteatoma, mastoiditis and meningitis [12].

Other autoimmune inflammatory diseases such as vasculitis, Henoch-Schönlein purpura and Kawasaki syndrome can also result in facial nerve palsy.

Rarely in children, facial nerve paralysis can be due to tumours such as schwannomas or hemangiomas of the seventh nerve or bone tumours such as rhabdomyosarcoma and histiocytosis. Pediatric facial nerve paralysis has been also described as an association with leukaemia [in many cases bilateral] or it could be due to parotid gland tumours [11].

Finally, traumas such as temporal bone fractures [longitudinal, transverse and oblique] can cause facial nerve palsy in children [13]. While iatrogenic paralysis can occur after surgery of the parotid gland, middle ear or mastoid [11,12].

Congenital facial nerve paralysis is an uncommon situation in newborns with an occurrence of 2 per 1000 live births. [18,19]. The major role of the physician is to differentiate between traumatic and developmental aetiologies. This distinction is important not only for the treatment options and prognosis but may also have medicolegal implications [1-5].

Developmental aetiologies include very rarely, isolated cases of aplasia/hypoplasia of facial nerve or their nuclei. There are very few cases of isolated congenital familial facial nerve agenesis/hypoplasia described in the literature. The first case of isolated complete congenital facial nerve agenesis was reported by Jervis et al in 2001 [20].

In our study we ,describe a child and his mother with congenital hereditary unilateral familial facial palsy. The child has another sibling and his dad who were both perfectly normal.

Conclusion

Pediatric facial nerve palsy is a condition with several implications, it causes significant concerns to parents due to the functional and aesthetic outcomes and it is challenging for doctors. The possible causes are many, however idiopathic facial paralysis, or Bell's palsy, is the most frequent form of facial paralysis in children. Congenital unilateral familial facial palsy is rare, but isolated congenital unilateral familial facial nerve palsy is even rarer. A careful diagnostic workout is always recommended, to establish the most appropriate treatment. Hopefully, in the future, regenerative medicine could offer new options for the treatment of this condition.

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