Congenital Facial Palsy with Bilateral Anotia

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ABSTRACT

Congenital facial palsy is generally considered developmental or acquired. Most of the cases are related to birth trauma. Herein we report a case of congenital facial palsy with bilateral anotia and external auditory canal atresia.

Keywords: Congenital facial palsy, anotia

Congenital facial palsy (CFP) is generally considered to be either developmental or acquired in origin. Developmental facial paralysis is associated with other anomalies including those of pinna and external auditory canal, ranging from mild defects to severe microtia and atresia. Herein we report a rare case of congenital right facial paralysis associated with bilateral anotia and atresia of right external auditory canal.

CASE REPORT

A 6-month-old male infant was admitted to the pediatric ward with lower respiratory tract infection. There was history of facial asymmetry and absent ears since birth. There was no history suggestive of intrauterine infection or drug intake during pregnancy. The baby was full-term normal vaginal delivery.

Physical examination showed bilateral anotia, preauricular tag was present bilaterally and right lower motor neuron type of facial palsy (Figs. 1-3). There was no other cranial nerve palsy and the rest of the examination including neurological examination was normal.

Magnetic resonance imaging (MRI) brain was normal. High-resolution CT temporal bone done to define the etiology of facial nerve palsy revealed absence of pinna, right auditory canal was not visualized and the middle ear ossicles were reported normal. Brainstem Evoked Response Audiometry (BERA) was normal.

DISCUSSION

Congenital facial nerve palsy is an infrequent condition with a reported incidence of 2.1 per 1,000 live births. In 78% of cases CFP is related to birth trauma. No such history was available in the index case. Other causes include, intrauterine posture, intrapartum...
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Multimodal Imaging of Pericardial Disease

- Acute pericarditis can be a practical issue in the US (STEMI mimics). Constrictive pericarditis is an often neglected disease.
- CCT, echo and cardiac MRI are the 3 diagnostic modalities.
- Size of pericardial space is directly proportional to the fluid. Rapidity of accumulation is also important and not just the amount of fluid.
- In a pericardial effusion, it is important to identify the amount of fluid, tamponade physiology and tissue characteristics.
- Timing of constriction when the mitral annulus velocity is >8 cm/s is especially in heart failure.
- CMR has a definitive role in diagnosis of a pericardial cyst: thin-walled, clear fluid and well defined margins.

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Some cases of CFP have been attributed to agenesis of the petrous portion of the temporal bone, with resulting agenesis of the facial and auditory nerves, the external ear and the mastoid region. Most commonly, development facial paralysis is associated with other anomalies. The most common site reported is the maxilla, including defects such as cleft palate, hypoplastic maxilla and duplication of the palate. Others have demonstrated a propensity for anomalies of the pinna and external auditory canal, ranging from mild defects to severe microtia and atresia. A striking association of grossly abnormal pinna, multiple defects and facial palsy has been reported in 9-15% of patients. The index case had bilateral anotia and right auditory canal atresia with right facial palsy.

Aural atresia occurs in approximately one in 20,000 live births. Atresia and microtia are parts of several syndromes with inherited defects or acquired embryopathies owing to intrauterine infections (rubella, syphilis), ischemic injury (hemifacial microsomia) or toxin exposure (thalidomide, isotretinon).

Embryonic insult, severe enough to cause aural atresia would also affect other organ systems. Aberration in the canalization process of external auditory canal can lead to stenosis, canal tortuosity or fibrosis/osseous obliteration. Since middle ear structure develops independently, the tympanic cavity and ossicles may be normal. Defects in the canalization process may also be associated with faulty formation of pinna. In the index case right side CFP was associated with anotia and right sided atresia. No other abnormalities were observed.

Several surgical techniques are employed for treatment of CFP. The ideal time for the intervention is controversial. Some clinicians advocate early (pre-school) surgery for the animation of children’s faces, while others propose surgery not before adolescence.

Muscle transplantation for facial paralysis has been shown to be effective. However, the possibilities of reconstructive surgery are limited. Traumatic facial palsy in neonates is associated with good prognosis. In contrast facial palsies, as in the index, case carry a poor functional outcome.
REFERENCES


Clues that may Suggest a Serious Underlying Cause of Hoarseness

Associated with hemoptysis, dysphagia, odynophagia, otalgia or airway compromise
Concomitant discovery of a neck mass
History of tobacco or alcohol use
Neurologic symptoms
Possible aspiration of a foreign body
Symptoms do not resolve after surgery (intubation or neck surgery)
Symptoms in a neonate
Symptoms in a person with an immunocompromising condition
Symptoms occur after trauma
Unexplained weight loss
Worsening symptoms