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Case Report

Case report of Anotia with ipsilateral facial nerve palsy

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ABSTRACT

Anotia is the severe form of microtia anomalies that involve pinna and external auditory canal present at birth is rare to be associated with congenital facial nerve palsy. The author reports a man with Right side Anotia and ipsilateral facial nerve palsy. This is rare report of such an association in adult patient had right Anotia and ipsilateral right facial palsy.

Keywords: Anotia, Facial nerve palsy, Microtia

INTRODUCTION

Congenital aural atresia (CAA) occurs in about 1 in 10,000-20,000 live births. Unilateral atresia is far more common than bilateral, on the order of 3-4:1. For unknown reasons, CAA occurs more frequently on the right side, and affects males more often than females. The CAA is marked by the absence or stenosis of the external ear canal, and is usually accompanied by the absence of a tympanic membrane, ossicular abnormalities, and microtia.[1-3] The primary embryologic abnormality in Congenital aural atresia CAA is failure of the first branchial groove epithelial plate to canalize.[4] Arrest of this process prior to canalization at 6 months gestation results in complete atresia (Anotia), and arrest during canalization results in incomplete atresia or canal stenosis. Because of the embryologic association between the external ear canal and the first branchial arch, patients with CAA often have a contracted middle ear space and ossicular abnormalities. Likewise, the first and second branchial arches give rise to the axonal hillocks that form the auricle, accounting for the common association between CAA and microtia. Conversely, the inner ear primarily derives from the otocyst rather than the branchial apparatus. As a result, inner ear structures are usually not affected in patients with CAA.[5,6] Microtia associated with facial nerve palsy, right side preponderance, and male predilection.[7] Facial palsy of developmental origin is associated with other anomalies including those of pinna and external auditory canal, which range from mild defects to severe microtia and atresia.[8]

Here, we report a 47 year-old male with Right side Anotia and right facial nerve paralysis.

CASE REPORT

A 47 years old male, was found to have right side Anotia with right side facial asymmetry. The patient was unable to close his right eye with the reduced blink of the right eye. There were no other congenital anomalies of eyes or limbs. On examination, he had right side Anotia, preauricular tags and atresia of Right external auditory canals (Figure 1). There was right lower motor neuron type facial palsy evident in the form of inability to close the right eye (Figure 2). A fistula of the second branchial groove also seen in this patient (Figure 1). Lipoidal injection into the fistulous track shows that it extends into the pharynx (Figure 3). There was no other cranial nerve palsy and the remainder of the neurological examination

was normal. Pure tone auditory show Rt side sever mixed hearing loss and normal left side.

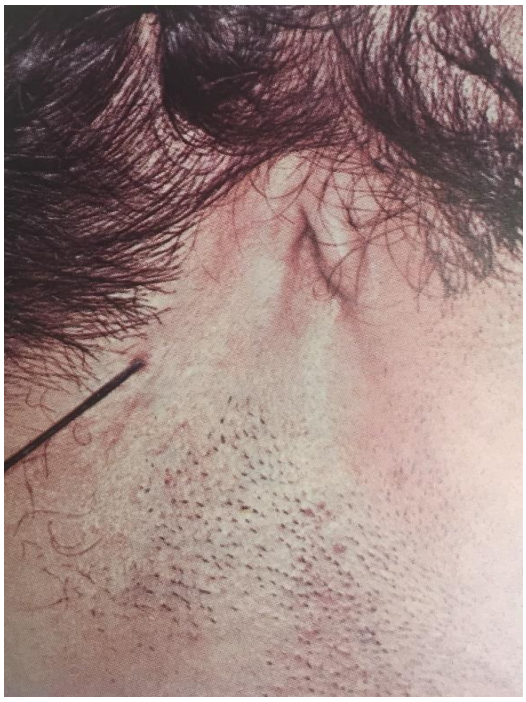

Figure 1: Anotia of right side with skin tags. External auditory meatus is not seen also a fistula of the second branchial groove seen at top of the black pointer.

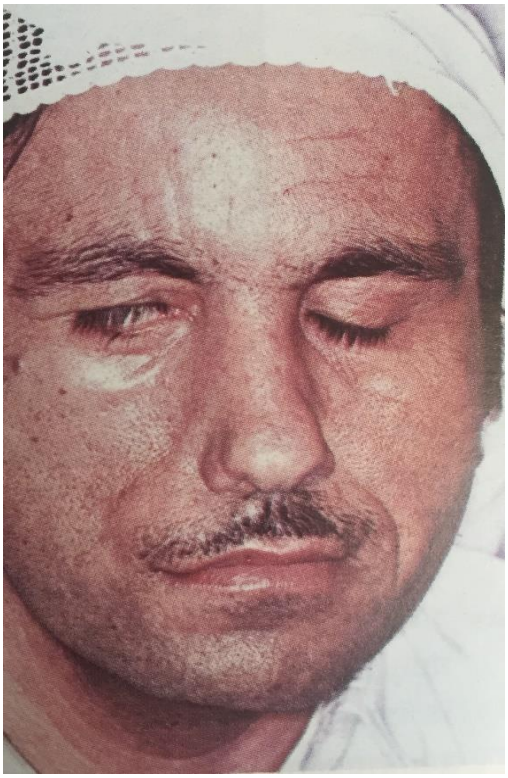

Figure 2: Lower motor facial palsy on the right side.

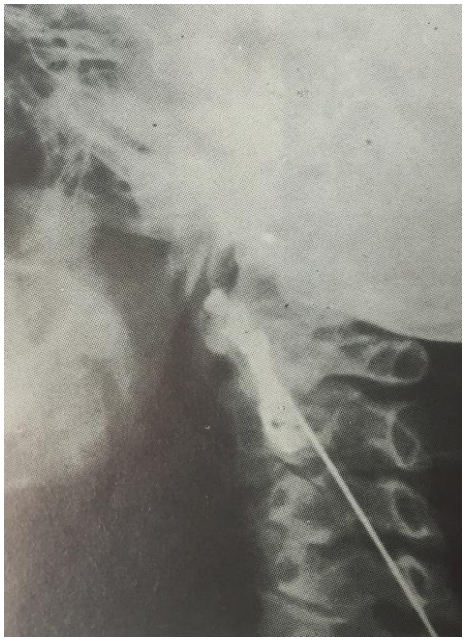

Figure 3: Lipoidal injection into the fistulous track shows that it extends into the pharynx also seen in this patient.

DISCUSSION

Congenital aplasia of the facial nerve nucleus is the most frequently reported etiology for bilateral congenital facial palsy. Syndromes such as Moebius, Poland's, and Goldenhaar's have CFP as part of their symptoms.[8] 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 have been described.[9] Developmental facial palsy is associated with abnormalities of the pinna and external auditory canal, ranging from milder defects to microtia and atresia. Microtia and atresia can be inherited as a part of several syndromes or, acquired due to intrauterine infections (rubella, syphilis), toxin exposure (thalidomide, Isotretinoin) or ischemic injury (hemifacial microsomia). A fault in the canalization process of external auditory canal leads to stenosis, canal tortuosity or fibrosis/osseous obliteration. Defects in the canalization process may also be associated with the faulty formation of the pinna. The development of middle ear structures occurs independently to that of the external ear.[10]

Hence, the tympanic cavity and ossicles may be normal. Gathwala *et al*, had reported a child with congenital right facial palsy associated with bilateral Anotia and atresia of the right external auditory canal. Pearl had described two girls with Anotia, facial paralysis and congenital heart disease without any obvious etiology.[11]

In the absence of other conditions, children with Anotia/microtia can develop normally and lead healthy lives as seen in our patient.

CONCLUSION

Anotia and microtia of pinna and external auditory canal atresia is rare syndromes to be associated with facial nerve palsy. Anotia and Microtia is mark for look for other congenital anomalies.

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