CASE REPORT

An Extremely Rare Case of Accessory Auricle, Polyotia and its Surgical Correction

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Abstract Certain malformations may occur during the embryonic development of the ear. Accessory auricle has a wide range from a small elevation of skin to a large size to resemble an additional auricle, where the latter is called polyotia. This condition is defined as an accessory auricle that is large enough to closely resemble an additional pinna rather than a skin remnant and cartilage. Polyotia, also known as mirror ear or accessory ear, is a type of ear anomaly in the tragus area, but this term refers to substantial anomalies which resemble an accessory ear, unlike a pre-auricular tag. Here, we present a case of an accessory auricle in a young female adult. The occurrence was unilateral; the subject did not have any audiological disorder. A simple surgical procedure is also mentioned for excision of accessory without any complications. We compared our findings with different cases of accessory auricle in the literature. The clinical importance and genetic association are also discussed.

Keywords Ear · Auricle · Accessory · Polyotia · Surgical correction

Introduction

Malformation of the external ear may occur alone or be a feature of congenital syndrome. The reported incidence is 1 in 12,500 births [1]. The association and proportions

between the structures in the face and head is a subject of interest for scientists as well as artists since renaissance. Auricle is one of these structures that have an important place in beauty measures, besides its function in hearing.

The external ear begins to develop around the dorsal end of the first branchial cleft during the sixth week of gestation. The auricle results from the fusion of six small buds of first two pharyngeal arches, called hillocks. The auricle is usually complete by the twelfth week. Initially, the auricles form at the base of the neck, but as the mandible develops, the auricles migrate to their normal adult location by gestational week 20 [2].

Accessory auricular anomaly is a cartilaginous skeleton covered with skin like a tragus [3]. It is frequently unilateral, but may be bilateral or multiple [4]. Embryologically, failure of proper fusion of six auricular hillocks during auricle development has been blamed for the development of accessory ears and polyotia [5].

Polyotia is an extremely rare congenital anomaly of the external auricle. This condition is defined as an accessory auricle that is large enough to closely resemble an additional pinna rather than a skin remnant and cartilage [6]. Polyotia, also known as mirror ear or accessory ear, is a type of ear anomaly in the tragus area, but this term refers to substantial anomalies which resemble an accessory ear, unlike a pre-auricular tag. We report a case in which the patient had a duplicated auricle that was corrected successfully.

Case Report

A 23-year-old girl reported to the Department of Oral and Maxillofacial Surgery, A.C.P.M. Dental College, Dhule, Maharashtra, India with a large accessory anomalous

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Fig. 1 Pre-operative

auricle on her left ear (Fig. 1). The accessory structure was approximately 3×2.0 cm in size and was composed of an elastic and fibrous cartilaginous component. This duplicated structure was positioned anteriorly to the original auricle. There was a cartilaginous concave bowl that resembled the conchal hollow of the human ear just behind this duplicated auricular structure. This accessory anomaly was not as large as the patient's external auricle, but it mirrored her left external auricle. One auricular canal existed between the two auricle-concha structures. Her family history did not reveal similar case of accessory auricle nor audiological disorder.

The surgical correction procedure used was very simple. The ultimate goal of the surgical procedure was to correct the tragus area. First, the photographs of the right ear were taken to know the accurate measurements and extent of the tragus. After taking the photographs, the reference points were marked over the accessory auricle region for proper excision and esthetics. Then the local anesthetic agent was injected in 1:100,000 concentration and the incision was given according to the reference points. After proper reflection of the skin and subcutaneous tissues, the cartilaginous part was exposed (Fig. 2). The cartilaginous part was then excised according to the reference points (Fig. 5). Then the excess skin was also trimmed off and the incision site was closed using 5–0 ethilon (Figs. 3, 4). There were no postoperative complications like infection over the operative site or facial nerve weakness and the patient was regularly recalled for follow up for 12 months (Fig. 6).

Discussion

There are several forms of developmental anomalies of the auricle including microtia, polyotia and accessory auricle. Microtia is a congenital anomaly characterized by a small,

Fig. 2 Intra operative

Fig. 3 Immediate excision

Fig. 4 Closure

abnormally shaped auricle (pinna). It is usually accompanied by a narrow, blocked or absent ear canal. Microtia can occur as the only clinical abnormality or as part of a syndrome [2].

Fig. 5 Excised auricle

Fig. 6 After 12 months

Polyotia is an extremely rare type of congenital external ear malformation. It is defined as an accessory ear that is large enough to resemble an additional pinna [7]. The aetiology of this condition is unclear [8].

Not more than thirty cases have been reported worldwide [7, 8]. Some cases were diagnosed with other congenital facial anomalies or craniofacial syndromes such as Goldenhar syndrome or Treacher-Collins syndrome [7, 8].

In addition, polyotia can present even in patients with normal ear conditions, but sometimes a constricted ear or a microtic ear can accompany [7]. In our case report, the patient presented with left polyotia and with bilateral prominent ears. Embryologically, the external part of the human ear is derived from first (mandibular) and second (hyoid) branchial arches. The tragus and the crus of helix are derived from the first branchial arch and other external structures are derived from the second branchial arch [9]. A recent etiology suggested that polyotia is caused by the

extraordinary migration of neural crest cells in the branchial arch during embryologic development.

Gore et al. [8] reported eight cases of polyotia and presented five salient points for surgical correction. The five points were: releasing the skin of the extra component, excision of extra cartilage, remnant skin trimming, preservation of facial nerve, and timing of operation. Previous reported surgical corrections for polyotia recommend excision of all accessory structures and using the excised cartilage to fill the accessory conchal hollow [7, 8]. In those cases, extra conchal hollows of polyotia were maintained as cartilage-linked cheek defects. The cheek defects were filled using excised accessory cartilages.

In our case, however, the patient only had a conchal hollow without a linked cheek defect and the hollow was small. Therefore, we decided to remove this hollow by collapsing the concha-resembling cartilage instead of filling the defect with excised cartilage. Because the shape and conditions of polyotia are very inconstant and the incidence of this anomaly is rare, it is difficult to establish a uniform surgical technique. The audiometric tests for hearing analysis and USG abdomen for renal function should also be performed to analyse the functioning of the organs.

The accessory auricular anomaly has been found in Goldenhar's syndrome. However, previous studies have shown that in many patients the accessory auricular anomaly is just an isolated symptom and that the genetic trait of accessory auricular anomaly is autosomal dominant [3].

Conclusion

Accessory auricle may be associated with certain syndromes. In such cases, complete audiological examination should be warranted, the subjects should be advised to receive genetic counseling for possible further examinations and surgical excision should be planned to achieve esthetic appearance of face.

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