Mirror Ear: A Rare Case of Polyotia

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ABSTRACT

Polyotia is an anomaly of the external auricle in which the accessory auricle 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 the term refers to substantial anomalies which resemble an accessory ear, unlike a pre-auricular tag. It is an extremely rare condition and to date, less than 30 cases of polyotia have been reported according to a review of the literature. A variety of theories has been proposed for development of such abnormal external auricle.

We report two cases of polyotia which presented to Dhulikhel Hospital ENT OPD that was successfully corrected surgically. An 8-year-old boy and a 14-year-old boy presented with a large accessory anomalous auricle on left and right ear respectively. The accessory auricle was composed of an elastic cartilaginous component covered with skin and was positioned anteriorly to the original auricle in both the cases. The anomaly was not as large as the patient's external auricle, rather it mirrored the external auricle. On the opposite ear both the cases presented with pre-auricular skin tag. Surgical correction was done by dissecting the skin free from the duplicated cartilage via an incision along the free edge of the helix the duplicated cartilage was contoured to fill the pre-tragal hollow and the tragus reconstructed with a free cartilage graft.

KEY WORDS

Accessory auricle, Accessory ear, Duplicated pinna, Mirror ear, Polyotia

INTRODUCTION

Polyotia is an extremely rare type of congenital external ear malformation and reported incidence is 1 in 12,500 births.[1] Marx defined polyotia as an accessory ear that is large enough to resemble an additional pinna.[2] The accessory auricular anomaly is a cartilaginous skeleton covered with skin like a tragus. It is frequently unilateral but may be bilateral or multiple.[3] The etiology of this condition is still unclear, and less than thirty cases have been reported worldwide. They can be associated with congenital facial anomalies or craniofacial syndromes such as Goldenhar syndrome or Treacher-Collins syndrome or can even present with normal ear conditions.[4,5]

Besides its function in hearing auricle has an important place in beauty measures. Embryologically, failure of proper fusion of six auricular hillocks during auricle development has been blamed for the development of accessory ears and polyotia.[3] Polyotia was first reported by Von Bol and De Kleyn in 1918, who described left polyotia in a child with multiple congenital anomalies.[6] The size and shape of accessory or anterior auricle was reported to be as large as the normal posterior auricle, and the size of the accessory auricle became the key clue in differentiating polyotia and skin tag. Posterior to the accessory auricle, the auricle in the correct position can be normal, constricted, or microtic. Because of this symmetry along the coronal plane between the anterior and posterior auricles, the condition is sometimes referred to as mirror ear.[4,6]

Many hypotheses have been proposed to explain the etiology of polyotia, with abnormal migration of neural crest cell (NCC) which is the most plausible of mechanisms. Retinoic acid embryopathy has also been known to cause embryological abnormalities, significantly affecting NCC migration. The patients with fetal exposure to retinoic acid had an increased risk of developing external ear defects including partial duplications.[7-9]

We report two different cases which presented to Dhulikhel Hospital ENT OPD with polyotia that was corrected successfully. An 8-year-old boy and a 14-year-old boy presented with a large accessory anomalous auricle on left and right ear respectively. The main components for surgical correction are deconstruction, cartilage grafting to fill out depressions, skin flap tenting over hollows and preservation of the facial nerve. These allow for good aesthetic reconstruction and thus enable more normal social function at this vulnerable age.

CASE REPORT

CASE 1

An 8-year-old male presented with a large accessory anomalous auricle on his left ear (Fig. 1a & b) and pre auricular skin tag on the right tragus (Fig. 2a & b) since birth. On examination, there was a 4.5X1.25 cm accessory anomalous auricle mirrored his left ear which has well-formed helix and conchal cavity composed of elastic cartilage. One atretic external auditory canal existed between the two auricle-concha structures. The helical crus of accessory ear were continuous with a normal posterior auricle, which had normal helix, antihelix, and antitragus. The left EAC measured about 2X2 mm. The pre-auricular tag on right tragus measured 2X1.5 cm with a normal auricle, which had normal helix, antihelix, and antitragus. On otoscopic examination, right tympanic membrane was pearly white, shiny, translucent, smooth with no bulging or retraction with cone of light in the anterior inferior quadrant. Despite the malformation, the left ear was positioned at the same level as the contralateral auricle, with anteroposterior view of the normal auricles being symmetric. Laboratory examination was within normal limits and pure tone audiometry showed 65 dB, 50 dB, 40 dB, 45 dB air-bone gap at 0.5, 1, 2 and 4 kHz on the left ear (moderate to severe conductive hearing loss) and normal hearing on right ear but moderate to severe conductive hearing loss in the left ear.

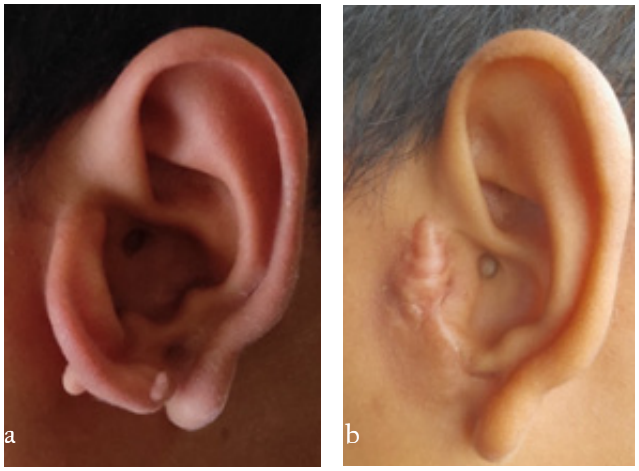

Figure 1a. Clinical photograph showing the accessory pinna in left ear with EAC stenosis and b. post-operative photograph 1 year after surgical correction of accessory pinna left ear.

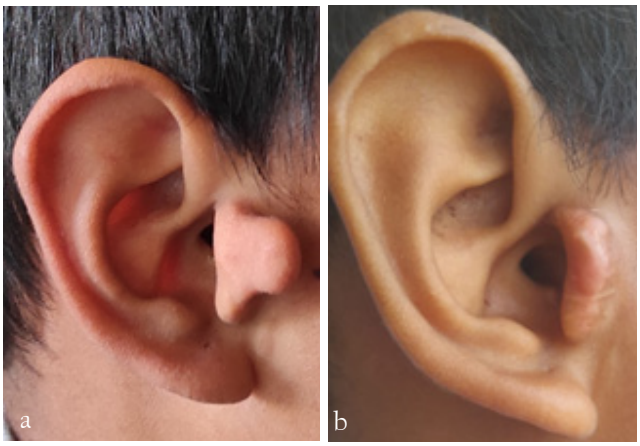

Figure 2a. Clinical photograph showing pre auricular tag in right ear and b. post-operative photograph 1 year after surgical correction of pre auricular tag in right ear

CASE 2

A 14-year-old male presented to Dhulikhel Hospital ENT OPD with a large accessory anomalous auricle on his right ear (Fig. 3a & b) and three pre auricular skin tag anterior to left tragus (Fig. 4a & b) since birth. On examination, there was a 4X1.5 cm accessory anomalous auricle located anterior to right tragus which is well formed helix and conchal cavity composed of elastic cartilage, with a depression that mirrored cavum concha along the longitudinal axis of external ear. The helical crus of accessory ear were continuous with a normal posterior auricle, which had normal helix, antihelix, and antitragus. The pre-auricular tag on left tragus measured 1X1 cm, 0.5X0.5 cm, 0.25X0.25 cm lying anterior to a normal auricle, which had normal helix, antihelix, tragus and antitragus. On otoscopic examination, bilateral tympanic membrane was pearly white, shiny, translucent, smooth with no bulging or retraction with cone of light in the anterior inferior quadrant. The right auricle was positioned at the same level as the contralateral auricle, with symmetrical anteroposterior view of the normal auricles. Laboratory examination and pure tone audiometry were within normal limits.

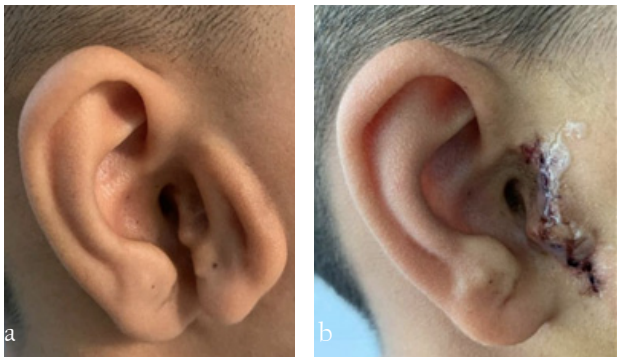

Figure 3a. Clinical photograph showing the accessory pinna in right ear and Figure b. post-operative photograph 7 days after surgical correction of accessory pinna right ear

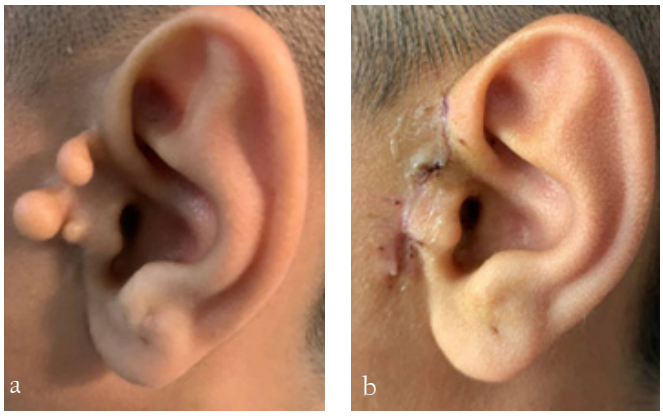

Figure 4a. Clinical photograph showing pre auricular tag in left ear and b. post-operative photograph 7 days after surgical correction of pre auricular tag in left ear

Surgical technique

Surgical techniques to correct polyotia have not been established because of its rarity and shape variation.[5] Gore et al. reported eight cases of polyotia and presented five salient points for surgical correction. These points were: releasing the skin of the extra component, excision of extra cartilage, remnant skin trimming, preservation of facial nerve, and timing of operation.[4]

Firstly, we photographed the mirrored and the opposite ear in both the cases 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 aesthetics. Under all aseptic condition the local anesthetic agent lignocaine 2% with adrenaline 1:200,000 concentration was infiltrated, and the incision was given according to the reference points. After proper reflection of the skin and subcutaneous tissues, the cartilaginous part was exposed. The overlying skin was released from the cartilage of the mirrored ear and excised according to the reference points. Cartilage sheets was pre-bonded with fine non-absorbable suture material (silk 3-0) to form a block for grafting to fill smaller non-confluent areas and contour tragus. The excess skin was trimmed off and the incision site was closed using prolene 3-0. The pre auricular tag on the opposite war was excised and closed in 1 layer with prolene 3-0 There were no postoperative complications like infection over the operative site or facial nerve weakness and the patient was regularly recalled for follow up.

During the hospital stay, the patient was given antibiotics to prevent postoperative infection and anti-inflammatory drugs postoperative pain and inflammation. Both the patients were discharged seven days after the operation with a good prognosis assessment. Facial nerve function was checked clinically by asking the patient to wrinkle the forehead to observe any asymmetry, close the eyes tightly to observe lid closure competence, smile to observe asymmetry of the nasolabial fold, and contract the mouth to observe orbicularis oris functions. There was no facial nerve dysfunction found during the examination.

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, 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.[4]

Polyotia is an extremely rare condition appearing sporadically in the surgical literature since the turn of the century and less than 30 cases have been reported.[4] The distinction of polyotia from a skin tag rests largely on size and a subjective level of resemblance to a normal external ear. The accessory pinna is not as large as the normal auricle, nor there is any complete duplication. While some are a mirror image i.e., the symmetry exists along the coronal plane between the anterior and posterior auricles, and others are based on a different axis.[4]

The etiology of this fascinating condition is unknown. Normal auricular development derives from the first (mandibular) and second (hyoid) branchial arches between the fifth and twelfth weeks of gestation. The auricle itself is formed from six 'hillocks', the three mandibular arch hillocks fuse to form the tragus and some surrounding pinna, whereas the three hillocks of the hyoid arch form the antihelix, the antitragus, the lobule and most of the helix. The external ear begins to develop around the dorsal end of the first branchial cleft during the sixth week of gestation.[4,12] The auricle results from the fusion of six small buds of first two pharyngeal arches, called hillocks and is usually complete by the twelfth week. Initially, the auricles form at the base of the neck, but migrate to their normal adult location by gestational week 20 as the mandible develops.[4]

Several etiologies of this condition have been proposed. Arey blamed failure of fusion of the first and second arch tissues for the development of accessory auricles and polyotia while Otto suggested in 1979 that they represent choristomas, derived from hyodidal ectoderm but developing in mandibular tissue.[10,11] However more evidence points to abnormal migration of neural crest cells causing polyotia. As early as the 1930s Harrison revealed that hindbrain grafts will induce the formation of an entire ear in estranged ectoderm in amblystoma punctatum. He proposed that this was due to neural tube tissue exerting an effect on facial ectodermal tissue.[4]

Similarly, the case described by Jackson et al. reported hindbrain duplication and polyotia present in the same case.[12] This parallels recent work demonstrating the effect of retinoic acid embryopathy (RAE). RAE has been known to cause embryological abnormalities for many years, but Wei et al. have shown in a primate model that deviation of cranial neural crest cells (NCCs) from their normal positional fate causes abnormal development of war.

Lammer et al. have proposed that RAE is also responsible for abnormal NCC migration during development in humans.[13] In a retrospective study he demonstrated that patients who had had fetal exposure to retinoic acid suffered an increased incidence of external ear defects including partial duplications. This evidence suggests a strong environmental influence on the etiology of polyotia. The number of cases in this series is too small to draw significant conclusions about the patient associations with the condition (sex, racial background, associated syndromes, etc.).[4,5,8-13]

The patients usually present early in a child's life but the surgery is best delayed until five years of age. At this stage the mirror ear can be removed before any major teasing at school occurs. The mirror ear often has a substantial conchal hollow and in these cases, exploration reveals a cartilage-lined cheek defect. This should be filled prior to skin closure with spare cartilage and may be achieved using excised cartilage from the mirror ear. Alternatively, the excess cartilage may be used to recreate a normal tragus if this is required. Cartilage sheets was pre-bonded with fine nonabsorbable suture material to form a block for grafting to fill smaller nonconfluent areas and contour tragus. Skin flaps may be used to improve the soft tissue contour as the Z-plasties where there is a potential contour abnormality in a grossly abnormal case.

The facial nerve develops at the same time as the external ear, and abnormalities of the course and/or size of the facial nerve might be expected to coexist. Concern about the position of the facial nerve or any duplicated part in relation to external ear duplication may warrant the use of a nerve stimulator. There is no role for attempting to image the facial nerve; imaging techniques are insensitive to branches of the facial nerve beyond its intra-parotid course. Preservation of any deep conchal cartilage of the mirror ear in the cheek may act as a shield to protect the facial nerve. Filling the resultant defect with spare cartilage graft and skin flaps is safer than excising deep cartilage remnants.

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