Polyotia, a rare occurrence-clinical presentation and surgical management: A case report from Abuja, Nigeria

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INTRODUCTION: Polyotia is a rare congenital condition that presents as a large accessory auricle, often mirroring the appearance of the normal auricle of the ear. The primary implication of this condition is in the esthetic challenges it poses to affected individuals. This article highlights our experience with the surgical management of a case encountered by our surgical foundation.

CASE REPORT: A nineteen (19) year old male student presented to our clinic at a surgical outreach program held at Kwali general hospital in July 2018 with a complaint of an abnormal looking left ear noticed since birth. The patient had concerns about his aesthetics and could not perceive sounds with the ear. A diagnosis of non-syndromic polyotia was made after detailed history and examination and successful surgical reconstruction to correct the deformity was done under local anesthesia.

CONCLUSION: We recommend early surgical reconstruction of polyotia to improve esthetics of affected patients.

Key Words: *Polyotia; Cleft and facial deformity foundation; Abuja; Nigeria*

INTRODUCTION

Polyotia, alternatively described as mirror ear, accessory auricle or duplicated auricle, is a congenital condition presenting as a large accessory auricle, often mirroring the normal auricle [1-4]. The accessory auricle is usually large enough to closely resemble the pinna unlike other anomalies of the ear which may present as an auricular tag or remnants of skin and cartilage. It may be unilateral or bilateral and may be associated with a syndrome [1,5]. Syndromes linked with polyotia include; Goldenhar syndrome [2-4], Treacher Collins syndrome [2-4], Brachmann-de-Lange syndrome [1], Wolf-Hirshorn syndrome [6], Townes-Brocks syndrome [6,7] and VACTERL syndrome [6]. The reported incidence is 1 in 12,500 live births [5]. The possibility of familial inheritance has been documented in the literature and has been notably associated with X-linked or autosomal recessive inheritance [6]. In addition, polyotia may be associated with the presence of an audiological disorder, mandibular and condylar hypoplasia [8]. The condition is very rare and about 30 cases have been reported according to Japati et al. [5].

The auricle develops around the dorsal end of the first branchial cleft during the sixth week of intra-uterine life (IUL) [5]. Fusion of the six small buds of the first two pharyngeal arches (hillocks) results in formation of the auricle, and this process is usually complete by the twelfth week of IUL [5,9]. The auricles initially form at the base of the neck but gradually migrate to their normal position following mandibular development. This process is usually complete by the twentieth week of IUL [5,9]. Failure of fusion of the six auricular hillocks during embryonic development has been postulated as the cause of polyotia [5,6].

The auricle is an important component of the face serving to project an individual's aesthetic features in addition to forming part of the audiological complex responsible for the special sense of hearing. It is the only visible part of the ear and functions like a funnel to direct sound into the middle ear *via* the tympanic membrane [10]. Females often adorn it with jewelries in order to enhance facial aesthetics. Thus, malformations or defects involving the auricle often impact negatively on the aesthetics and self-confidence of sufferers as it may be a source of ridicule in young individuals as well as stigmata for the older persons. Surgical correction of these deformities ultimately improves facial aesthetics which eventually translates in a better quality of life for affected individuals [5]. We present a young male with a unilateral accessory left auricle which was surgically corrected successfully.

CASE REPORT

A nineteen (19) year old male student presented at the 20th Cleft and Facial Deformity Foundation (CFDF) surgical outreach program held at Kwali General Hospital, Abuja, Nigeria in July 2018 with a complaint of an abnormal looking left ear that had been present since birth (Figure 1a). History revealed that the patient had concerns about his facial aesthetics and his inability to hear sounds with the ear. He further reported no positive family history of similar occurrences or any other audiological disorder.

Examination of the patient revealed a left accessory auricle which was approximately 3.5 2 cm in size located in the pre-tragal region, anterior to the primary auricle and composed of skin and cartilaginous components. The accessory auricle had a well formed conchal cavity and helix with a conchal depression along the longitudinal axis. There was a depression on the face anterior to the accessory auricle with associated hyperpigmentation of the skin. The helical crus of accessory ear was continuous with the primary left auricle which was also malformed (with the fusion of the tragus and anti-tragus) resulting in the absence of a well-defined tragus, a miniature anti-tragus and the absence of an inter-tragic notch. An obliteration of the left external auditory canal was observed, with skin completely covering the orifice.

On the right side of the face, there was a skin tag anterior to the tragus of the right auricle which measured about 1 × 1 mm (Figure 1b) although the right auricle appeared normal. The lower jaw and facial midline were deviated to the left and the patient had a prominent left antegonial notch (Figure 1c). Examination of other systems revealed no significant findings.

Investigation conducted included an orthopantomogram (OPG) and a pure tone audiology test. The OPG confirmed mandibular hypoplasia on the left with a prominent left antegonial notch (Figure 1d). Pure tone audiological evaluation revealed a moderately severe mixed hearing loss more on high tones on the left while the right ear was normal following the audiological test. A diagnosis of non-syndromic polyotia of the left auricle, and a right pre-auricular skin tag were made. The patient was planned for surgical excision of the accessory auricle and reconstruction of the left ear to be carried out under local anaesthesia.

SURGICAL PROCEDURE

Treatment goal was to excise the accessory auricle and to reconstruct the original auricle to restore a near normal aesthetic appearance with emphasis

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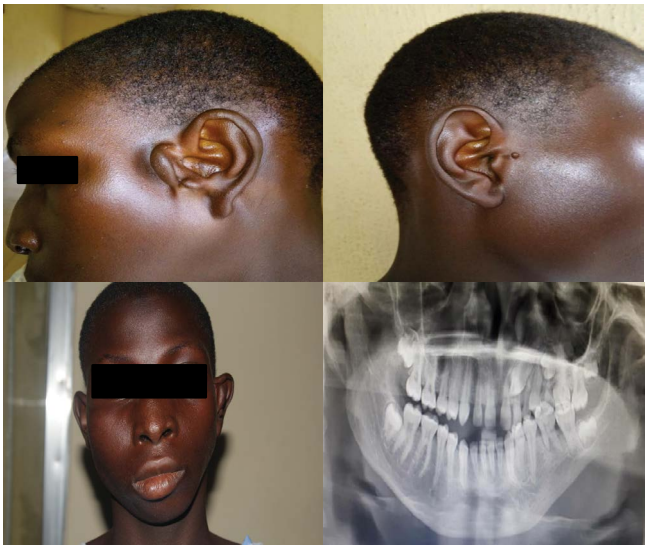

Figure 1) *(a) Left ear malformation in a 19 year old boy with presence of an accessory auricle; (b)Presence of a normal auricle on the right with a skin tag in the pre-tragal region; (c) Left Mandibular hypoplasia with a prominent antegonial notch and deviation of the jaw to the left; (d) Orthopantomograph showing left mandibular hypoplasia*

on reconstruction of the left tragus and the left inter-tragic notch. Pre-operative photographs of the normal auricle on the right was taken and used as a guide for reconstruction of the left auricle.

Informed consent was obtained from the patient [11] and he was placed supine on the operating table. Skin preparation and draping was done in the standard fashion after which markings for the skin incision and appropriate reference points were made with a sterile surgical pen. Local anaesthesia of the auricle and surrounding preauricular and postauricular regions were achieved using 2% lignocaine hydrochloride containing 1:100,000 adrenaline with a 7 minute wait period observed to enable adequate vasoconstrictive effect. Incision was made through skin and subcutaneous tissue anteriorly and posteriorly to excise the attachment of the accessory auricle to the face and blunt dissection was done to expose the cartilage on the accessory auricle (Figure 2). The cartilage and excess skin was then excised, and haemostasis was achieved promptly. Further, a 1 cm vertical incision was made through the skin, subcutaneous tissue and cartilage to separate the fused tragus and anti-tragus and to recreate an inter-tragic notch. Haemostasis was achieved and the wound edges were sutured with 4/0 nylon sutures (Figure 3a) and a compressive gauze dressing placed on the ear.

Post-operatively, medications administered included antibiotics (Amoxicillin 500 mg, 8 hourly for 5 days) and analgesics (Diclofenac Potassium 50 mg, 8 hourly for 3 days). No complications were recorded and the patient was satisfied with his appearance at the second month follow-up visit (Figure 3b).

DISCUSSION

Polyotia is described as the presence of an accessory auricle closely resembling an additional pinna rather than a skin remnant or cartilage [4]. The aetiology of accessory auricle is not known [3]. However, retinoic acid embryopathy (RAE) has been recently implicated in the pathogenesis of polyotia in cases of fetal exposure to Accutane (isotretinoic acid) which results in abnormal migration of neural crest cells in the branchial arch during embryogenesis and hindbrain duplication [12,13] The history of polyotia can be traced to von Bol et al. [14], who first reported the case in 1918. Although less than 30 cases of this congenital deformity have reported in the literature [5], none have emanated or focused on a predominantly black population. In fact, a PubMed and HINARI search of polyotia in Africans returned no entry, and this underscores the importance of reporting this case. Our patient is male and even though the auricular defect was observed on the left, affectation of the left and right sides of the face has been reported [5,8].

The patient under review is a 19-year-old male who presented at an outreach program that offered free surgical services to patients across the country. A complaint of an abnormal left ear noticed since birth was made by the patient. The delayed presentation by the patient was as a result of his inability to access specialist healthcare facilities which is a common problem in underserved communities in most African countries.

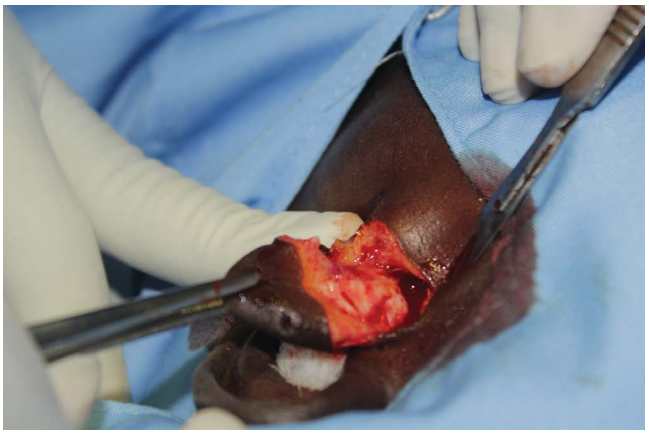

Figure 2) *Incision through skin and subcutaneous tissue with excision of the accessory auricular cartilage*

Figure 3) *(a) Immediate post-operative outcome; (b) 2 months post-operative review*

The clinical features observed in our patient are similar to reported cases [1,3-6,8]. Japati et al. [5] reported a similar case of a patient who had a normal right ear with an accessory auricle on the left mirroring the auricle. This is also similar to the reported case by Gadre et al. [1] and Moon and Oh [3]. In contrast however, the reported cases by Japati et al. [5] and Moon and Oh [3] had patent external auditory canals with normal hearing unlike our case report where the external auditory canal on the affected side was completely sealed by skin with associated hearing impairment. Moon and Oh [3] also reported the presence of hemifacial microsomia in addition to the accessory auricle in their patient. Gadre et al. [1] reported a syndromic association with polyotia with the patient having mental retardation, stunted physical growth, limited speech, a micrognathic mandible, shallow glenoid fossa with absence of pneumatization of both mastoids. They thus made a diagnosis of polyotia in association with Brachmann-de-Lange syndrome.

The aetiology of the accessory ear could not be identified which is in keeping with the idiopathic nature of polyotia [3]. The embryological origin of the external part of the human ear is the first branchial arch (mandibular arch) and the second branchial arch (hyoid arch) [15]. Therefore, an abnormality involving the arches will affect the structures derived from them. This theory explains the presence of mandibular hypoplasia along with the external ear abnormality observed in our patient. The obliterated ear canal is responsible for the hearing impairment seen in our review. Impairment

in hearing is a consistent finding in patients with polyotia [2,3,16], and this was observed in our patient from the history findings which was confirmed with pure tone audiometry. Our case is consistent with previous reports with respect to the clinical features vis a vis the presence of an accessory auricle anterior to the prominent pinna, a skin tag anterior to the contralateral ear, a cartilaginous concave bowl that resembled the conchal hollow of the human ear located just posterior to the duplicated auricular structure and left-sided mandibular hypoplasia [3,8]. Regarding the assessment of associated mandibular asymmetry in individuals with polyotia, Rha et al. [8] employed computed tomography (CT) scans in their work, although in this case, an orthopanthomogram (OPG) was employed for the same purpose. The orthopantomogram offered several advantages in this setting as it was readily available, inexpensive with a lower ionizing radiation received considering the indication for the investigation and gives a good view of the mandible from one mandibular condyle to the other which allows for instant comparison of both sides of the mandible.

Surgical excision of the accessory auricle and reconstruction of the affected ear has been documented by previous investigators as the treatment of choice [2,8], although no consensus exists concerning the most appropriate surgical technique for reconstruction of polyotia [3]. Goodman et al. [17] advocated surgical excision of the anomalous cartilage and lobule, with reconstruction of the tragus, which was also supported by Gore et al. [2] where they summarized five points that aid in the surgical correction of polyotia. These include-release of skin of the accessory auricle, excision of accessory cartilage, trimming of skin remnant, preservation of facial nerve and appropriate operation time. In our case, the goal of reconstruction was to improve facial aesthetics and restoration of self-esteem. The reconstruction of the auricle was achieved through surgical excision of the accessory cartilage with careful preservation of the branches of the facial nerve and apposition of the wound margins with a non-resorbable suture material under local anaesthesia. This method of reconstruction is the same as that reported by Goodman et al. [17] and Japati et al. [5] where the accessory auricular cartilage was excised. However, this method is in slight contrast to the technique by Gore et al. [2] where they utilized the excised auricular cartilage to fill the depression anterior to the accessory auricle. Moon and Oh [3] employed a two stage approach to surgical management unlike our case report where a single stage approach was adopted. In their report, the aim of the first surgery was to correct the tragus area, while the second surgery was done later to eliminate the concha resembling cartilaginous bowl. While they reported good post-operative results with their approach, the rationale for staging the procedure was not reported by the authors. Our surgical approach to treatment involved a single stage procedure with reconstruction of the inter-tragic notch on the affected ear after surgical excision of the accessory auricle in contrast to reports by Moon and Oh [3] and Japati et al. [5] where the inter-tragic notch was not reconstructed. It is the opinion of the authors that the reconstruction of the inter-tragic notch contributes to restoring the auricle to its normal appearance. Despite the difference in technique highlighted by different authors in the literature [2-4,8,17], they have all been reported to have a favorable outcome with subsequent patient satisfaction. Overall, the important denominator observed in these various techniques reported, including ours, is improved facial aesthetics and restoration of self-esteem following surgical reconstruction.

CONCLUSION

Polyotia is a rare, and when it does occur, it may be psychologically challenging due to its potential negative effects on facial aesthetics, self-esteem and function. However, the role of surgical reconstruction in restoration of facial aesthetics and self-esteem has been documented to be successful at achieving these aims, even to near normal levels. In this light, we recommend early surgical reconstruction in order to prevent these deleterious sequelae in affected individual.

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