รายงานผู้ป่วย / Case Report
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Accessory auricle: A case report and review of the literature

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Abstract

Accessory auricle is a congenital anomaly of a pinna which is commonly localized anterior to the tragus. It contains a small piece of cartilage underneath. The prevalence of accessory auricle is 0.2-0.47%. It can be found isolated or related with otherassociated disorders. Excision is usually indicated for cosmetic reason. Here, we present a case of isolated accessory auricle and review of the literature, including the embryological development of pinna and a genetic - linkage analysis.

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รายงานผู้ป่วยภาวะติ่งเนื้อหน้าใบหูแต่กำเนิด และบทความทบทวนวรรณกรรม

กชพร วงษ์สุวรรณ
สาขาวิชาโสต ศอ นาสิกวิทยา ศูนย์การแพทย์ปัญญานันทภิกขุ ชลประทาน
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บทคัดย่อ

Accessory auricle เป็นความผิดปกติแต่กำเนิดของใบหูมีลักษณะเป็นติ่งที่มีส่วนของกระดูกอ่อนอยู่ภายใน ซึ่งตำแหน่งที่พบบ่อยคือส่วนหน้าของ tragus ความชุกของความผิดปกตินี้เป็นร้อยละ 0.2-0.47 อาจพบความผิดปกตินี้ได้เพียงอย่างเดียวหรือพบร่วมกับความผิดปกติของอวัยวะอื่นๆ ร่วมด้วย ในที่นี้เรานำเสนอกรณีศึกษาผู้ป่วยที่มี accessory auricle โดยไม่มีความผิดปกติของอวัยวะอื่นๆ พร้อมกับการทบทวนวรรณกรรมรวมถึงการพัฒนาของใบหูของภาวะตัวอ่อนในครรภ์และผลการวิเคราะห์ความเชื่อมโยงทางพันธุกรรม

Introduction

The diagnosis of accessory auricle may be missed because of its infrequent occurrence. Accessory auricle is an elastic cartilaginous skeleton covered with skin like a tragus. It is most commonly localized just anterior to the tragus or ascending crus of the helix. If the condition is large enough to resemble an additional pinna, it will be defined as polyotia, an extremely rare condition, which only twenty cases had been reported in the world literature[1]. It differs from the preauricular tag which does not contain a cartilage underneath[2].

The external ear is derived from the first (mandible) and second (hyoid) branchial arches. The tragus and the crus of helix are mainly formed by the first brachial arch, while the helix, antihelix, superior crus, inferior crus, and earlobe are evolved from the second branchial arch[1-5]. The accessory auricle is an anomaly caused by embryologically failure of proper fusion of the first branchial arch during auricle development. The treatment of accessory auricle is surgical removal for cosmetic. Since this condition can related to other associated disorders, a complete examination and audiological examination should be considered. This article is purposely for review and updates the related abnormality with accessory auricle.

Case report

A 19 year-old man presented at birth with an accessory auricle, approximately 15 mm. in diameter, located anterior to the right tragus. He had no other abnormality, and had no either familial history of accessory auricle nor head and neck deformity. Physical examinations revealed a soft tissue with cartilage underneath located just anterior to the

right tragus with well - formed helix, external auditory canal, and tympanic membrane (Figure 1). Rinne and Weber test by using tuning fork 512 Hz were performed, and the test were within normal. Surgical excision under local anesthesia was discussed for cosmetic purpose. A longitudinal elliptical incision line was made through skin and subcutaneous tissue. The underlying cartilage was excised. The skin was closed meticulously (Figure 2-4). Our patient, however, did not have any abnormal symptoms. He was satisfied with the results of surgery.

Discussion

A pinna is formed during embryonic life from six tiny hillocks, at the dorsal end of the first (mandible) and second (hyoid) branchial arch.Failure of fusion at the fourth week of gestation causes of accessory auricles or polyotia. The first hillock forms the tragus, the second and third hillocks form the anterior crus of the helix while the fourth and fifth

Illustrations

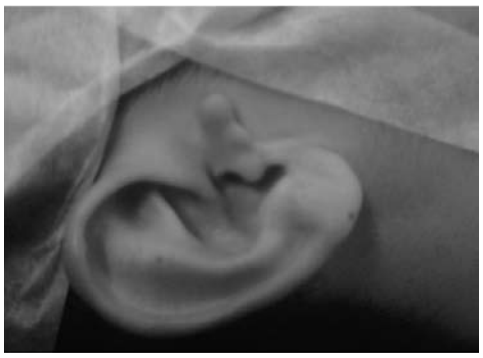

Figure 1 There was a large accessory auricle anterior to a tragus.

Figure 2 The right preauricular area was prepped and draped properly. After 1% xylocaine with 1:100,000 epinephrine was infiltrated, a longitudinal elliptical incision line was made with the # 15 blade. The incision was made through skin and subcutaneous tissue.

Figure 3 The underlying cartilage was excised and bleeding was stopped. The skin was closed meticulously with nylon suture 5-0.

Figure 4 The specimen of an accessory auricle, the cartilage covers with normal appearance skin.

hillocks form rest of the helix and antihelical crura, and finally the sixth hillock become the antitragus. However, the external auditory canal occurs after complete canalization of the mesoderm from the first branchial arch. Failure of this process results in a narrow, blocked or absent ear canal (aural atresia or stenosis). The auricle and the cartilaginous external auditory

canal are derived from the same origins during the same period in embryonic life. Therefore, the anomaly of auricle and external auditory canal are usually occur together[1-4]. The auricle is usually completely formed at the twelfth week and as the development of mandible, the auricles migrate to their normal location by the twentieth week[5].

Accessory auricle is a small excrescence of skin like a tragus that contains an island of elastic cartilage and most commonly localized just anterior to the tragus or ascending crus of the helix[5, 6]. It's frequently unilateral, but may be bilateral or multiple [3,7]. Small cartilage underneath makes it differ from the preauricular tag which can be fall off at early ages if it pedunculated. The typical sizes of the excrescences are 3-10 mm. If it has large size resembles, so an additional auricle, it will be defined as a rare condition[3]. Accessory auricle may relates with abnormalities of the tragus, preauricular fistula, and neurological disorder[8]. Accessory auricle may be found in Goldenhar's syndrome called oculoauriculovertebral dysplasia (OAV). It is an incomplete development of the ear, nose, soft palate, lip, and mandible affected on only one side of the face. It associates with anomalous development of the first and second branchial arch. Common clinicalmanifestations include limbal dermoids, preauricular skin tags, strabismus and vertebral malformations. Other associated disorders include Wolf-Hirschhorn syndrome (microcephaly, micrognathia, short philtrum, prominent glabella, ocular hypertelorism, dysplastic ears, and periauricular tags), Treacher-Collins syndrome (hypoplasia of the facial bones, ear anomalies, eye problems, and cleft palate), Townes-Brocks syndrome (abnormalities of the external ears, anorectal malformations, renal abnormalities, heart abnormalities, hand and foot abnormalities) and VACTERL association (vertebral defects, anal defects, cardiac defects, esophageal defects, renal defects, limb defects, and growth defects). The prevalence of accessory auricle is 0.2-0.47%[3, 8, 9]. Without sex differentiation[6]. An isolation of accessory auricle is a genetic trait of autosomal dominant with complete penetrance. Yang et al[6], reported a genetic- linkage analysis in the isolated symptom of accessory auricular anomaly with microsatellite markers spanning of the whole human genome in the Chinese family with 11 affected individuals[6]. Linkage analysis defined the isolated ADAAA locus to a 9.84 cM interval between markers D14S283 and D14S297at chromosome 14q11.2-12. However, the accessory auricle is also valuable in the alar reconstruction. Lin et al[10], have used the accessory auricle as a composite island flap to reconstruct alar defect in patient with basal cell epithelioma of nasal alar by microvascular

anastomosing the superficial temporal vessels to the facial vessels with excellent outcome.

Conclusion

Accessory auricle can be found alone or related with other anomalous developmental syndrome. Therefore, the complete physical examination including audiological tests and familial history taking is recommended. The treatment is excision for cosmetic purpose.

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