

## Case Report

# Multiple accessory tragi in a male child

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### ABSTRACT

Accessory tragus (AT) is a congenital abnormality of the external ear which was first described by Birkett in 1858. AT is typically found in the preauricular region, though it can also occur less commonly in the mandibular or cervical regions. This cartilaginous protrusion, commonly known as the preauricular tag, is a rudimentary tag of ear tissue. With an incidence rate of 0.2-0.5%, this abnormality is believed to be an autosomal dominant hereditary condition caused by an abnormality in the development of first or second branchial arch. Patients with accessory tragus are more likely to be males and typically appear unilaterally. It may exist alone or in groups with a distinct morphology and location. Here, we report a case of multiple accessory tragi in a male child affecting the tragus and cheek with no other malformations.

**Keywords:** Accessory tragus, Branchial arch, Tragus, Skin tag, Preauricular region, Cheeks

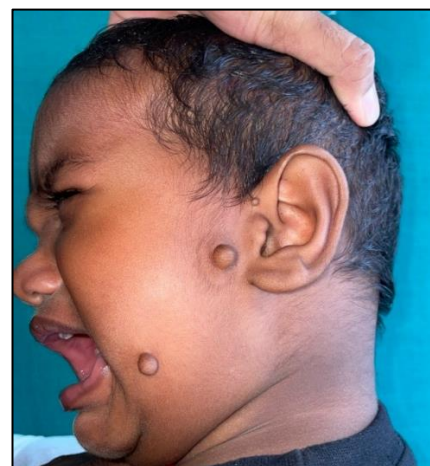
### INTRODUCTION

Accessory tragus (AT) is a relatively common congenital anomaly that may be observed during routine examination. Single or multiple accessory tragus may occur along a line connecting the tragus of ear and oral commissure, mainly at the tragus, preauricular region and cheek. Similar lesions in the neck have been termed as “cervical auricles” or “cervical skin tags”. Accessory auricles of the preauricular region and cervical auricles differ developmentally. Hence, the term “cervical chondro-cutaneous branchial remnants (CCBRs)” is proposed for these abnormalities. Only a few reports describe these lesions.<sup>1</sup> AT are mostly benign, cause no functional impairment and are usually excised mainly for cosmetic reasons, with majority of them consisting of elastic cartilaginous tissue on histopathological examination (HPE).

### CASE REPORT

A male child which was one year old had several growths on his left side of the face, between his tragus and angle

of mouth, present since birth. A tag was seen above the tragus in front of the left ear and two nodules between the left ear and angle of mouth.



**Figure 1: Multiple well-defined skin-coloured nodules present over an imaginary line joining from ascending crux of helix of left ear to the angle of mouth.**

The child had no other congenital malformations. On dermatological examination, the left ear showed two nodules that were about 1 cm in diameter extending from the tragus to the angle of the mouth and a skin-coloured, hard, non-tender tag that was 3 mm in size seen anterior to the crux of helix (Figure 1). Parents did not mention any additional physical abnormalities or known medical conditions. The child was examined by ophthalmologists and otorhinolaryngologists which revealed no other abnormalities. Echocardiography and abdominal ultrasonography were both normal. There is no similar history in the family members. The parents refused to undergo a skin biopsy. The patient is under follow-up.

## DISCUSSION

The prevalence of AT is approximately 0.2%. Embryologically, the auricle develops from the first pharyngeal arch, which originates at the oral commissure. The predilection site of accessory tragus is along an imaginary line connecting the tragus and oral commissure in the path of migration of first branchial arch. This line coincides with the junction of the maxillary and mandibular prominences, which are derived from the first branchial arch. The most frequent localization is on the face in front of the tragus. Accessory tragus affecting the cheek usually co-occurs with similar lesions over the preauricular region. In rare instances, it occurs in post-auricular region and along the anterior border of sternocleidomastoid muscle.<sup>2</sup> It may be a single deformity or associated with other conditions such as cleft palate or lip, mandibular hypoplasia and abnormalities of the eyes and spine.

Multiple accessory tragi is associated with many congenital syndromes such as Goldenhar's syndrome (oculo-auriculo-vertebral dysplasia), VACTERYL syndrome, Wolf-Hirschhorn syndrome, Treacher-Collins syndrome, Townes-Brocks syndrome, Delleman syndrome (oculo-cerebro-cutaneous syndrome), Haberland syndrome (encephalo-cranio-cutaneous lipomatosis), Wildervanck syndrome, Nager acrofacial dysostosis and Down syndrome.<sup>3-5</sup> Goldenhar syndrome is characterised by a triad of accessory tragi, mandibular hypoplasia and ocular (epibulbar) dermoids.<sup>6</sup>

Treacher-Collins syndrome is characterised by facial dysmorphism, hypoplastic zygomatic bones, small mandible and eye abnormalities. Townes-Brocks syndrome is associated with multiple exostoses in addition to ear anomalies. Delleman syndrome is characterised by orbital cysts, microphthalmia and CNS malformations. Haberland syndrome is characterised by nevus lipomatosis, subcutaneous lipomas and focal skin aplasia. Wolf-Hirschhorn syndrome is characterised by facial dysmorphism, intellectual disability, growth retardation, hypotonia and congenital heart defects. In our case detailed examination and investigations were done to rule out syndromic associations. Although familial occurrences of AT have been documented in the past, it is

usually sporadic. According to previous studies, cardiac abnormalities, sensory neural hearing loss and renal anomalies are also associated with accessory tragus.<sup>7,8</sup> Excisional biopsy revealing vellus hair follicles, elastic cartilage, central fibroadipose tissue and prominent connective tissue framework in subcutaneous fat confirms the diagnosis of accessory tragus.<sup>9,10</sup> The differential diagnosis includes, Acrochordon (skin tag) which presents as a soft, skin coloured or hyperpigmented papule, usually located on the neck, axillae or periorbital region. Histology shows polypoid growth of loose to dense collagenous stroma with thin-walled dilated blood vessels in the centre. Adnexal tumour is flesh-coloured papules arising from adnexal structures of skin such as sebaceous glands, sweat glands, apocrine glands or hair follicles. Auricular fistula, also known as preauricular cyst or congenital auricular fistula is a congenital defect which presents as a cystic nodule or an invagination in the preauricular area. Histology shows a pit or cyst lined by stratified squamous epithelium. Cartilage is not present. Branchial cleft cyst presents as a cyst in the preauricular area, mandibular region or along the anterior border of the sternocleidomastoid muscle, most commonly presents in the second or third decade. Histology shows a cyst lined by stratified squamous or pseudostratified ciliated columnar epithelium.

Epidermoid cyst presents as a mobile, dermal nodule or cyst with a central punctum. Histology shows a cystic cavity filled with laminated keratin lined by stratified squamous epithelium. Hair follicle nevus presents as a small dome shaped papule from which fine hairs protrude evenly from the surface, commonly located on the face, in the vicinity of the ear. Histology shows closely arranged normal vellus hair follicles and absence of cartilage in the subcutaneous fat. Lipoma presents as a round to oval, soft, mobile subcutaneous nodule with a normal overlying epidermis that may be located in the subcutaneous fat. Fibroma or fibroepithelial polyp presents as a small, firm nodule with a smooth surface. HPE is characterised by hypocellular fibrovascular connective tissue that lacks cutaneous adnexal structures and vellus hair.

## CONCLUSION

In our case, a one-year-old male child presented with three accessory tragi along the line joining the tragus of ear and angle of mouth without any other congenital anomalies and syndromes that were previously documented in association with accessory tragi. Accessory auricle can be found isolated or associated with other developmental anomalies and syndromes. Therefore, complete physical examination including audiological tests and familial history taking is essential. The treatment is excision.

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