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Accessory Tragus: A Possible Sign of Goldenhar Syndrome

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The accessory tragus is a relatively common benign congenital anomaly. The tragus is a cartilaginous projection that normally occurs anterior to the external auditory meatus. Although aberrancy of the tragus may occur in isolation and is exclusively derived from the first branchial arch, it may occasionally signal a defect in the first or second branchial arches. Thus it may be a sign of other syndromes, such as oculoauricularvertebral dysplasia (Goldenhar syndrome). In fact, accessory tragus is a constant feature of this syndrome and may be associated with other syndromes. Accessory tragi are polypoid and should be distinguished from acrochordon (skin tags), as the shave excision commonly employed for skin tags may expose cartilage and cause slow healing or chondrodermatitis nodularis chronica helices.

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The accessory tragus is a relatively common congenital malformation. Although it usually occurs in isolation, it may occasionally signal a defect in the first or second branchial arches and reflect a variety of syndromes, most commonly oculoauricularvertebral dysplasia (Goldenhar syndrome). The accessory tragus needs to be distinguished from an acrochordon (skin tag), as treatment options differ.

Embryology

The pinna, or external portion of the ear, arises from the first 2 branchial arches, which are located near

the first branchial groove. Six tubercles or hillocks form on each arch: 3 from the first branchial (or mandibular) arch, and 3 from the second branchial (or hyoid) arch. During the first, fourth, and fifth weeks of embryonic development, these arches migrate superiorly with the growth of the embryo.¹⁻³ The first hillock has 2 portions; the ventral portion contributes to the mandible while the dorsal portion forms the tragus,⁴ which is the only portion of the ear forming in totality from the first branchial arch. The remaining portion of the pinna is a mixture of mandibular and hyoid origin but is mostly derived from the second and not the first branchial arch. Further evidence that the origin of the tragus is from the first branchial arch alone is supported by cases of agnathia,¹ which displays an absence of the mandible as well as portions of the internal ear and tragus, with the rest of the external ear appearing well-formed.

Clinical Features

Accessory tragi are flesh-colored, unilateral papules or nodules. They can be sessile or pedunculated, occur alone or with others, range in consistency from fleshy and soft to cartilaginous, and are frequently covered in vellus hair (Figure).⁴ There may be one large accessory tragus and several smaller satellite ones. Normally the tragus occurs at the anterior portion of the external acoustic meatus and juts backward. An accessory tragus may occur anywhere in the migratory path of the first 2 branchial arches. Usually papules or nodules occur in the preauricular region on or near the tragus, but occasionally they may occur elsewhere, such as on a line extending from the corner of the mouth to the ear, in the submandibular area near the angle of the mandible, or anterior to the sternocleidomastoid on the neck.⁴ Cartilage is an inconsistent feature and may not be evident on physical examination.⁵ The accessory tragus primarily is a clinical diagnosis based on time of onset and

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A flesh-colored accessory tragus covered in vellus hair.

anatomic location. Bilateral, nonfamilial, isolated accessory tragi have been described.⁶ Accessory tragi may be discovered when evaluating an infant with skin tags and no other abnormalities.

Pathology

Accessory tragi tend to be polypoid with a thin stratum corneum and many irregularly spaced vellus hair follicles. A deeper layer normally consists of a central core of elastic cartilage surrounded by a fibrous capsule. Two components that are invariably seen are an extensive connective tissue network in subcutaneous fat and numerous follicles of varying placement, which are associated with sebaceous glands.^{4,6,7} There may be aggregates of mature adipose tissue, a central plate of elastic cartilage, and sometimes small amounts of skeletal muscle. Histologic features of a hair follicle nevus may coexist.⁸

Epidemiology

It is estimated that accessory tragus can occur as frequently as in 1 to 2 births per 1000,⁹ though to our

knowledge no large studies have been done to assess prevalence. Isolated cases are much more common than those linked with other developmental abnormalities; the incidence of hearing loss and difficulty with speech production are thought to be higher in these isolated cases.¹⁰ One study showed that 5.8% (3/52) of cases are associated with developmental abnormalities,⁴ usually with facial malformations of the first branchial arch, such as a cleft lip, cleft palate, or hypoplasia of the mandible; the incidence of these malformations increases the closer the tragus is located to the mouth.

Of more consequence are associations with several major syndromes, all with first and second branchial arch malformations. Oculoauriculovertebral dysplasia, also known as Goldenhar syndrome, is an autosomal-recessive, phenotypically variable syndrome consisting of malformations in the first and second branchial arches; the accessory tragus has the most consistent relationship with these syndromes. Goldenhar syndrome is diagnosed most commonly as a triad consisting of accessory tragi, epibulbar dermoids or lipodermoids, and auricular fistula, and also may be associated with deafness.^{11,12} Mandibulofacial dysostosis, or Treacher Collins syndrome, is an autosomal-dominant condition most often consisting of ocular defects, external ear malformations, and hypoplasia of the face. It has a more irregular association with accessory tragi. Other syndromes occasionally associated with accessory tragi include otomandibular dysostosis, Townes-Brocks syndrome, VACTERL syndrome, Wolf-Hirschhorn syndrome, and Delleman syndrome.

Differential Diagnosis

Other conditions commonly confused with accessory tragi include acrochordon (skin tag), branchial cyst, auricular fistula, epidermoid cyst, cutaneous cartilaginous rests, hair follicle nevi, congenital midline hamartoma, and lipoma.

Treatment

Accessory tragi may be of sufficient size to produce cosmetic concerns. Although accessory tragi have been eliminated by simply tying twine or string around the base and waiting for them to fall off,⁵ they should be excised. Usually they cannot be removed by shave excision, as this process exposes cartilage and may cause slow healing or chondrodermatitis nodularis chronica helices. The treatment of choice is to make a small elliptical excision and remove all cartilage with smooth cutting of the cartilage.¹³ Superficial exploration and removal may be desirable to avoid other nearby deeper structures; however, most often

the accessory tragus is posterior to branches of the facial nerve and superior to the parotid gland and its duct.

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