CASE REPORT

Accessory Tragus Associated with Other Alterations. A Visual Compilation of Observed Cases and A Brief Review of the Subject

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ABSTRACT

The accessory tragus is a relatively uncommon congenital anomaly of the first branchial arch. Although there are reports of association with other pathologies, congenital or not, these are very rare. These associations are reviewed and some cases of the practice are presented in a very succinct way to show the existence of these associations. As this is a visual display of mostly congenital associations, medical records are not displayed.

INTRODUCTION

Accessory Tragus (AT) is a congenital anomaly of the first branchial arch. It is described as a cartilaginous projection anterior to the external opening of the ear. More specifically, it is defined as a formation, sometimes pedunculated, covered by skin, located in front of the tragus, as unilateral or bilateral, single or multiple, sessile or pedunculated (Fig 1a and 1b), soft or firm, the latter indicates the absence or presence of underlying cartilage. The solitary form, presenting as papule or nodule, is the most commonly observed.[1]

Despite the multiple existent reports on this malformation, articles whose focus is the association with other congenital pathologies, are relatively scarce. In

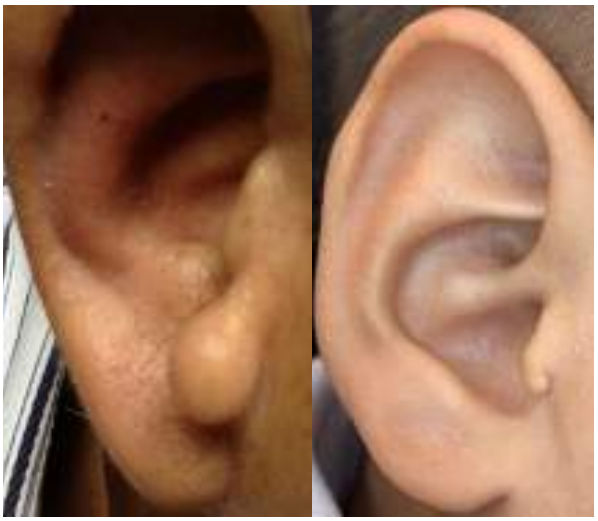

Figure 1a. Pedunculated AT. *Figure 1b. Non-pedunculated AT.*

his article, Al Aboud[2] indicates that although syndromes that occur with AT are rare, four syndromes were indeed found: Delleman, Goldenhar, Haberland and Townes-Brocks, whose highly summarized characteristics are discussed later on. Bahrani's work[1] adds three more syndromes: the Treacher Collins syndrome, VACTERL and Wolf Hirschhron syndrome. Rankin et al's AT report,[3] associated with Goldenhar syndrome, refers to a study that shows 5.8% (3/52) of AT cases are associated with developmental abnormalities, generally facial malformations of the first branchial arch, such as cleft lip, cleft palate or hypoplasia of the jaw. These malformations increase the closer the tragus is to the mouth. It has been reported that the presence of auricular lesions may be associated with kidney abnormalities, hearing loss, and cardiac abnormalities.[4]

Malformations of the external and middle ear are classified in:[6]

1. Malformations of the pinna.
    a. Mild or grade 1 auricular malformation (the ear is smaller with well-defined parts)
    b. Moderate or grade 2 auricular malformation (structural abnormalities, such as absence of the earlobe, helix, tragus abnormalities, etc.)
    c. Severe or grade III auricular malformation (total loss of the auricular structure with rudimentary appearance presenting with cutaneous-cartilaginous fold).
    d. Grade IV auricular malformation (anotia or absence of the pinna)

2. Malformations of the external and middle ear canal.
    a. Major malformations (absence of the ear canal and tympanic membrane (atresia), among other alterations.
    b. Minor malformations (normal or somewhat stenotic external ear canal with conductive hearing loss).

FIRST CASE

Microtia associated with AT

Incomplete development and growth of the pinna can lead to a small, deformed (microtia) or absent (anotia) organ[5] which can present as an isolated process, or associated with genetic syndromes or other malformations. Among such clinical pictures, the ones who stand out are Treacher Collins syndrome in children, hemifacial microsomia or Goldenhar syndrome, Townes Brocks syndrome, imperforate anus, dysplastic pavilions, thumb malformations, and cardiac and/or kidney involvement.[7] The ear canal is often narrowed or missing, leading to significant hearing decrease. Although there are several classifications of microtia, the most common form (grade 3) presents absence or severe malformation of most of the pinna. Only the earlobe is recognizable.[8] This case presents a small and partially developed left auricle. Absence of the external auditory canal and presence of preauricular tumor corresponding to single pedunculated accessory tragus (white circle). On the right side, the presence of AT is also seen (Fig. 3).

SECOND CASE

Agenesis of external ear canal associated with AT

Aplasia and hypoplasia of the external ear canal are rare otorhinolaryngological alterations, characterized by defects in the development of the external ear canal, which results in the presence of different malformations, varying from complete absence to mild stenosis and middle ear malformations, being generally unilateral and presenting deafness while compromising the pinna, external ear canal and middle or bilateral ear.[9] The pa-

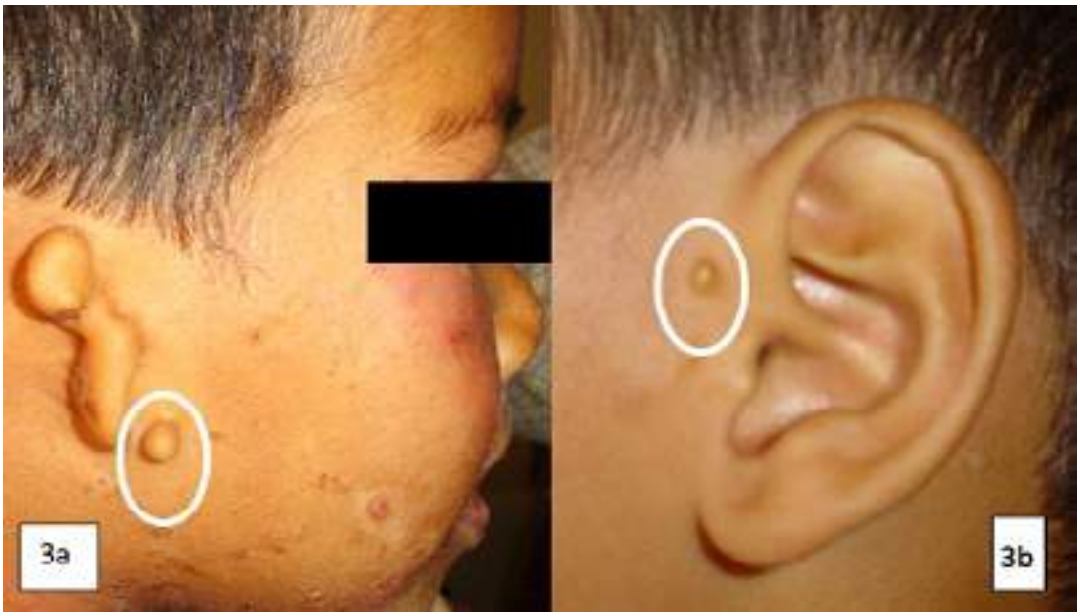

Figure 3a. First Case. Microtia and agenesis of the external ear canal associated with pedunculated AT (white circle).
Figure 3b. Left preauricular accessory tragus (white circle).

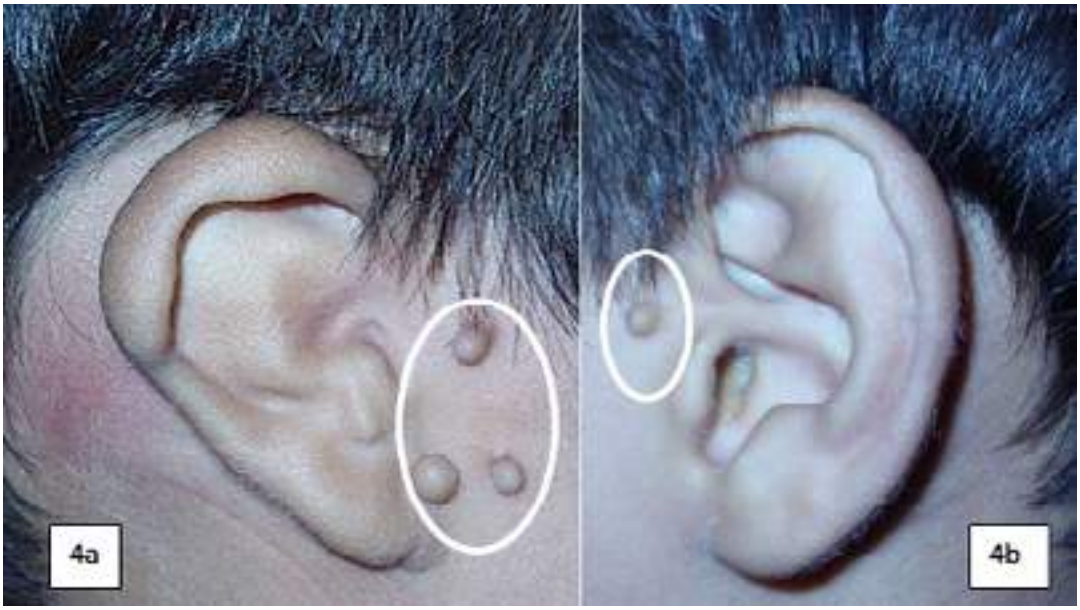

Figure 4a. Second Case. External auditory canal atresia associated with multiple right-sided AT (white circle).
Figure 4b. Left preauricular accessory tragus (white circle).

tient manifests absence of the antihelix and its anterior and posterior branches, as well as absence of the triangular and scaphoid fossa and total absence of the external auditory canal on the left side. Likewise, there are three pedunculated lesions corresponding to multiple AT. Presence of preauricular AT on the right side. (Figures 4a and 4b)

THIRD AND FOURTH CASES

AT associated with congenital fistulas

First described by Van Heusinger in 1864, the preauricular fistula, also known as sinus, fossa, tract or prearicular cyst, is a benign congenital malformation of preauricular soft tissues. Global estimates of its in-

cidence range from 0.1 to 0.9%. No symptoms are observed. It is considered as a physical examination finding due to infection and discharge. Usually unilateral, rarely bilateral, manifesting often in women more than men.[10]

In general, there are two classic forms pertaining to the clinical picture, the preauricular and a variant acknowledged as postauricular, which in turn is subdivided into three subtypes according to location (crus or shell).[11] Another classification that Bofill cites and that seems to be appropriate for most cases is the one that classifies them according to their location and embryological origin into three groups:[12]

1. Preauricular fistulas between the corner of the mouth and the tragus. (Fourth Case)
2. Preauricular fistulas located in front of the ascending hellix and leading to the external auditory canal (Third Case)
3. Sacciform depressions or small blind fistulas that can be located anywhere in the pinna.

The third case (Fig 5) presents non-pedunculated AT accompanied by a congenital preauricular fistula, classified as Bofill type 2, while the fourth case presents with multiple pedunculated AT in a 5-month-old baby girl. The latter is accompanied by a fistula located between the commissure and the tragus (type 1). Presenting hemangioma on the back.

FIFTH CASE

Multiple AT associated with earlobe malformation

The earlobe has two well-defined anatomical portions, considered as pendulous or nonpendulous, if it only has the cephalic portion, one is the cephalic portion (attached to the skull) and the other is the caudal portion (free). There are different earlobe shapes, the most common being round, square and triangular. However, there are forms of elongated earlobes pulled towards the cheeks, called pixie ears, with loss of pendulous contour; when acquired it is due to rhytidectomy with excessive traction of the area.[13]

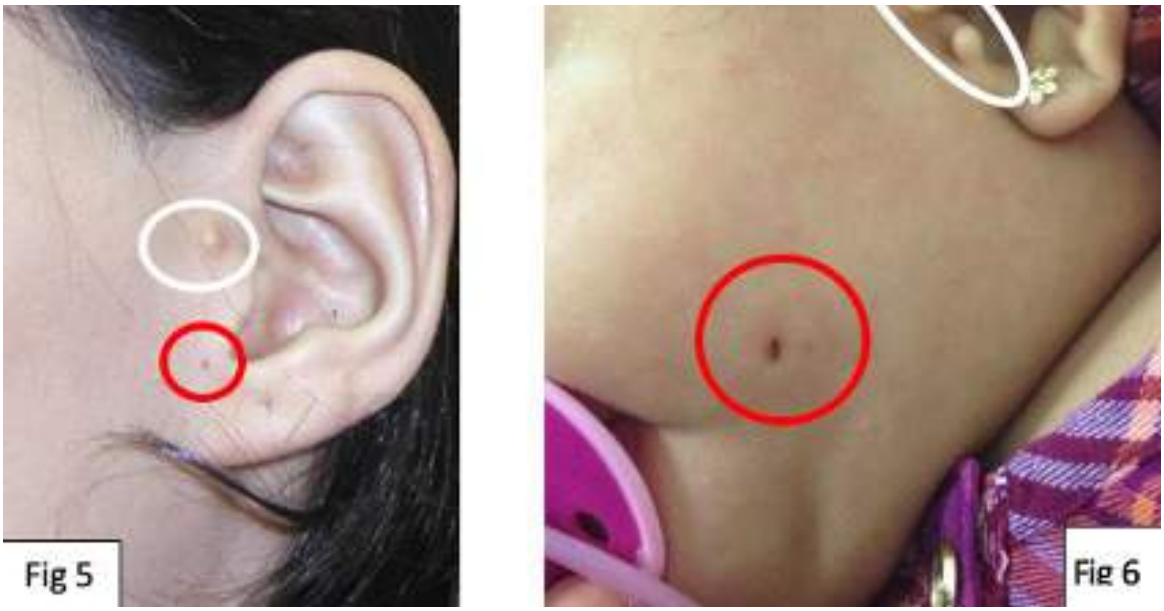

Figure 5. Third case. Type 2 congenital preauricular fistula (red circle) associated with AT (white circle).
Figure 6. Fourth case. Type 1 congenital preauricular fistula (red circle) associated with multiple AT (white circle).

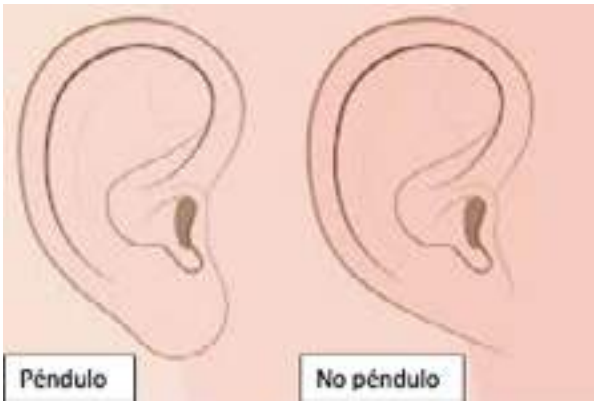

Figure 7. Difference between pendulum earlobe, free (left) and non-pendulum, glued (right).
Extracted from Google images.

The fifth case (Fig 5) exemplifies the presence of multiple preauricular AT, observed in the pavilion on the left side, associated with malformation of the earlobe, elongated towards the cheek, causing loss of the classic pendulous characteristic. Low pinna implantation is also observed.

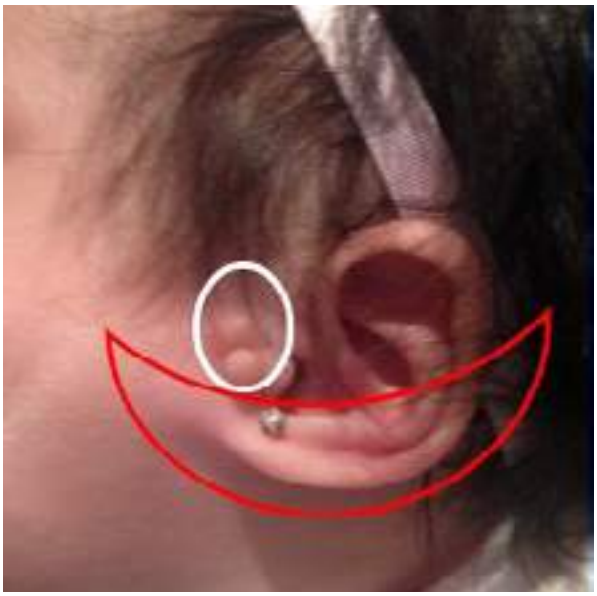

Figure 5. Fifth Case: Presence of multiple AT (white circle), associated with malformation with very prolonged earlobe, nonpendulum, pulled towards the cheek (red border).

SIXTH CASE

AT associated with localized left hemifacial flattening

This case has encountered findings that, except for the presence of the second AT, have been difficult to identify. The smallest is a classic pedunculated tragus and the second is larger with an inverted horn-like shape. The ear is thickened and flattened in the vertical axis. Localized facial flattening is observed in front of the tragus. The presence of periorbital edema is observed. All lesions are on the right side, the left is normal. (Fig 6a) The characteristics observed in this case are somewhat reminiscent of hemifacial microsomia or Goldenhar syndrome or oculo-auriculo-vertebral dysplasia, which some authors such as Gorlin and Pindborg believe is a variant of the Craniofacial microsomia, whose clinical characteristics include flattening of the face on the affected side due to maxillary and malar hypoplasia.[14] External ear malformation can range from complete aplasia to a distorted and wrinkled pinna.[15] As observed, the flattening of the ear, the AT and facial flattening follow an imaginary line that goes from the aforementioned structures towards the labial commissure (Fig 6b).

Figure 6a y 6b. Sixth Case. Multiple ATs (white circle) and a localized hemifacial flattening (red circle) are observed. The latter follows a line (red arrow) that goes from ATs towards the commissure of the mouth. The pinna is distorted.

SEVENTH CASE

AT within the Treacher Collins syndrome

The presence of AT can occur within multiple genetic and syndromic processes. The best known are named down below.

- Goldenhar syndrome (Oculo-atrial-vertebral dysplasia)
- Treacher-Collins Syndrome (Mandibulofacial Dysostosis)
- Townes-Brocks syndrome
- 4P minus syndrome
- Delleman Syndrome (DS Oculocerebrocutaneous)
- Familial Turner Syndrome
- VACTERL Association

Among them, the one that concerns us in this patient is the Treacher-Collins Syndrome (mandibulofacial dysostosis) which presents with the following alterations:

- **Auricles:** external ear eural atresia , abnormalities of the bones of the middle ear.
- **Craniofacial:** cleft or high-arched palate, choanal atresia, molar defects, mandibualr and zygomatic hypoplasia, temporomandibular joint disorder-glossoptosis, hypertelorism, micrognathia, nasal deformity.[16]

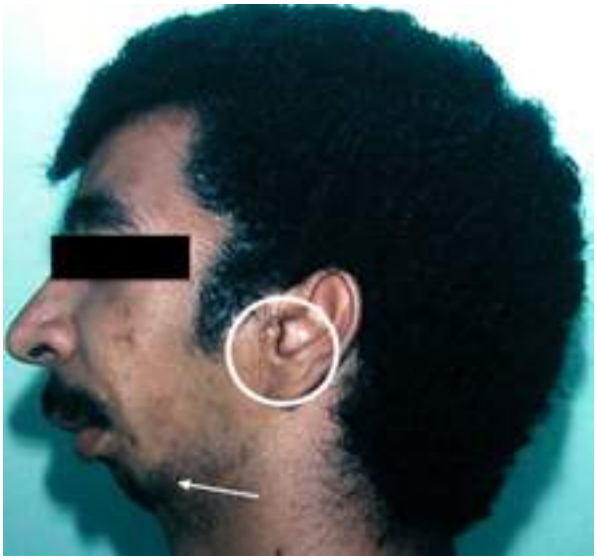

Figure 7. Treacher Collins syndrome associated with AT (white circle).

At the time of consultation, the patient had AT of the left ear and mandibular hypoplasia marked with the characteristic bird's face. He revealed that he had already undergone surgery once, as shown by the scar that can be seen in the mandibular arch.

The main characteristics of the clinical picture are outlined in the graphic (Fig 8)

Figure 8. Schematic graphic of the Treacher Collins syndrome.

EIGHTH CASE

AT associated with Vitiligo and Down Syndrome

In this regard, there are already reports on the association of AT with Down syndrome,[17] which is why this potential correlation should be taken into account. Regarding the association with Vitiligo, only the report by Gaukar et al. has been found. It reports the case of a patient with Goldenhar syndrome who presented with AT and vitiligo.[18]

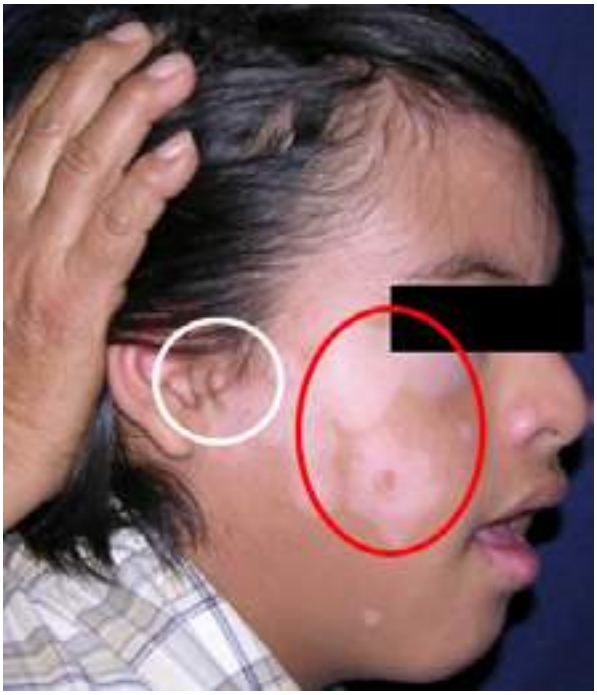

Figure 9. AT (white circle) associated with Down syndrome and vitiligo (red circle).

NINETH CASE

AT associated with relapsing Polychondritis

Relapsing Polychondritis is an inflammatory disease of recurrent appearance within the connective tissue. Preferential location pertains to the pinna, nose and the Tracheobronchial tree. An association of this condition has been previously reported in this patient.[19]

TENTH AND ELEVENTH CASE

AT associated with elongated earlobes

Although these two cases do not correspond to syndromic or genetic images, their relevance is due to the chance of presentation and association with AT.

In general, the length of the lobe should not exceed 30% of the total length of the ear. Over the years, the atrial lobe suffers elongation. At 60 years old, people's lobes expand a third of their length at age 20. This is partly attributed to gravitational force and redundant facial skin, occurring with age. The condition is more frequent in women than in men. Earrings are a source of additional weight for the ears and can become a factor which increases their length.[20]

Case ten (Fig. 11a) presents a patient suffering from tear of the ears due to earring theft, which changed the structure of the lobe and apparently contributed to its elongation. In case eleven (Fig 11b), an elderly patient shows ear elongation, probably related to age and constant use of earrings. Both cases present with classic AT.

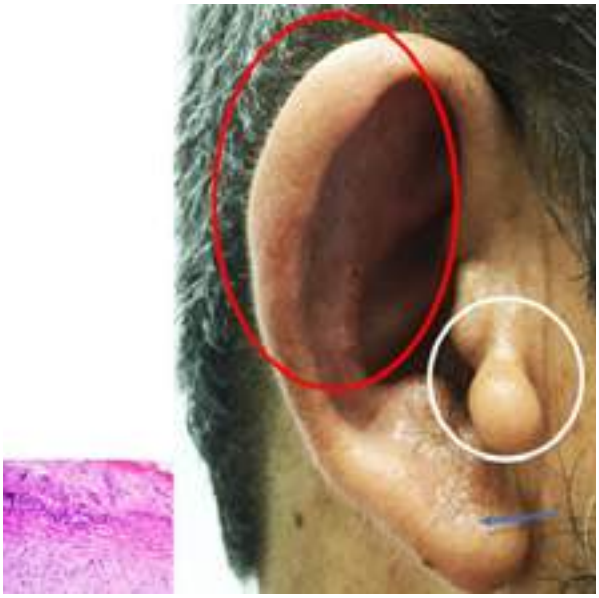

Figure 10. Recurrent Polychondritis + AT. Erythema and inflammation of the right pinna (red circle). Sparing of earlobe (blue arrow). Presence of pedunculated tumour arising from the anterior area of the tragus (white circle). Pathology shows the dermis with superficial perivascular infiltrate and capillary dilation.

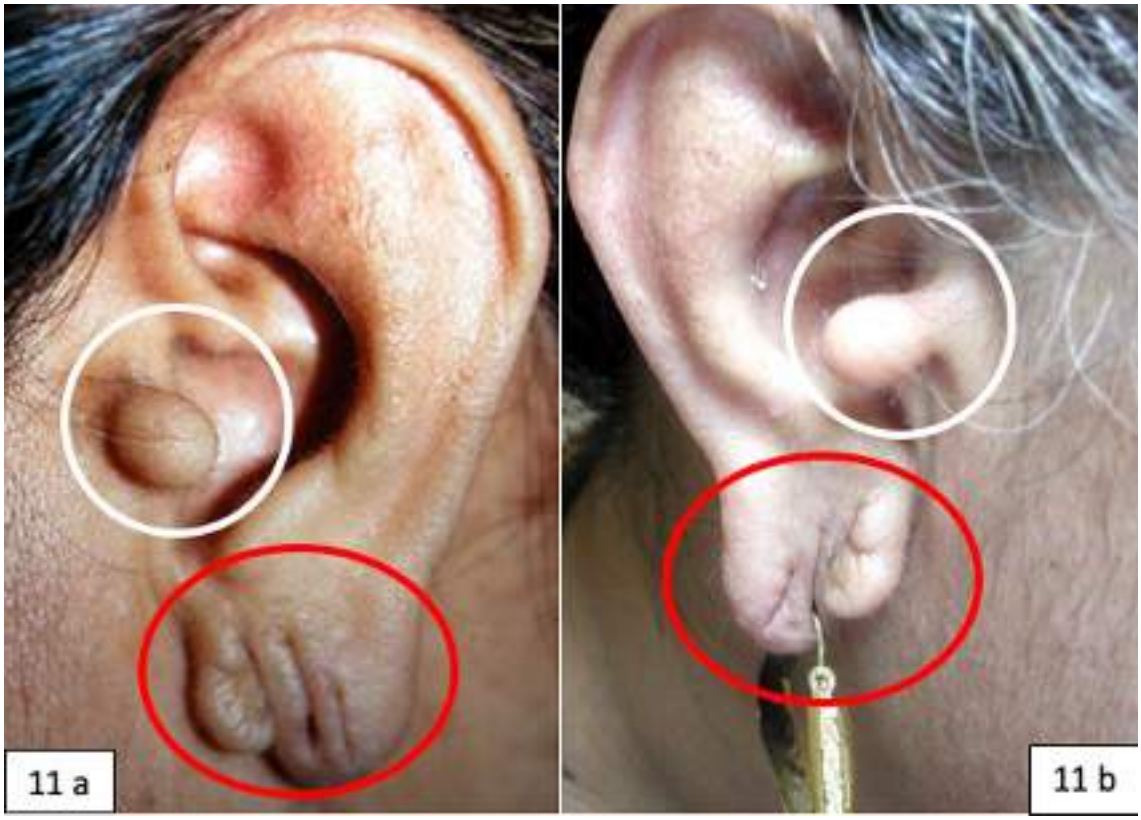

Figure 11a. Tenth Case: AT (white circle) and association with torn elongated earlobe (red circle).
Figure 11b. Eleventh Case: AT (white circle) and association with elongated earlobe because of age and jewlery (red circle).

CONCLUSION

AT is a malformation that manifests alone or in conjunction with other pathologies. It is also possible to find it associated with other genetic processes and syndromes of which it is already a common component. It is imperative to remember that its presence can be an indicator which leads us to investigate concomitant pathologies.

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