CASE REPORT

Relapsing polychondritis associated with accessory tragus. Case report and brief review of the literature

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ABSTRACT

Relapsing polychondritis is an inflammatory disease of recurrent appearance of the connective tissue, infrequently observed and preferentially located in the pinna, nose and tracheobronchial tree. The accessory tragus is defined as congenital supernumerary auricular structures considered as an anatomical curiosity, which corresponds to an anomaly of the first branchial arch, present from birth, and may contain eccrine glands, pilosebaceous units and cartilage.

We present the case of a 66-year-old patient who consulted for presenting an inflammatory process, accentuated, bilateral, of auricular pavilions with few days of evolution but with recurrent character, although in the previous episodes the process was unilateral. The condition presented for the first time 7 years earlier. In the region of the tragus of the right auricular appendage there was a tumor of pedunculated appearance of about 2 cm in diameter that the patient recalled had been present all his life and that corresponded to a unilateral accessory tragus. Histopathological study of the inflamed area is performed and a brief bibliographic review of both subjects is made.

INTRODUCTION

The pathology of the ears corresponds to one of those somewhat forgotten chapters of dermatology that can be specific to the area, or be part of a generalized picture or a complication or extension of other processes.

Relapsing polychondritis (RP) is an inflammatory disease of repetitive appearance of the conjunctive tissue, infrequently observed and preferentially located in all cartilaginous structures such as the pinna, nose and tracheobronchial tree, chest wall, peripheral joints and also in other areas rich in proteoglycans such as eyes, middle ear, kidney, heart and blood vessels.[1-2-3]

This entity was described by Jaksch-Wartenhost in Prague in 1923 who called it polychondropathy, then the disease received different denominations as systemic chondromalacia, panchondritis, chronic atrophic polychondritis and rheumatoid chondritis, to finally Pearson et al. in 1960 gave it its definitive and more appropriate name as PR,[4] being considered as a multisystemic, chronic disease, of a probable autoimmune origin[3] and with an estimated incidence of 3.5 cases per million inhabitants and year.

The Accessory Tragus (AT) is an entity that is bibliographically cited with a great variety of names such as poliotia, accessory auricle, supernumerary ear, branchial appendage or preauricular appendage among many others. They are congenital supernumerary auricular structures considered as an anatomical curiosity that correspond to an anomaly of the first branchial arch and that, being a malformation is always present from birth, although many times it is not taken into account. It may contain eccrine glands, pilosebaceous units and cartilage. Its origin can be hereditary and family cases are reported, but it can also be simply embryopathic.[5]

CLINICAL CASE

A 66-year-old male patient with controlled hypertension and a history of drug allergy to penicillin. One son diagnosed with xeroderma pigmentosum.

The patient consults for presenting erythema and edema of the auricular region, bilateral, accompanied by moderate pain on both sides, but predominantly on the left side. The erythema and swelling are present on the earlobes (Fig. 1 and 2). The pinnae are edematous and with a marked increase in volume (Fig. 3 and 4). On the right ear, arising from the anterior part of the tragus, there is a tumor with a pedunculated appearance, normal skin color, with a shape resembling a drop-shaped earring, which clearly corresponds to the diagnosis of AT.

The patient refers that the picture dates back to seven years before when he presented a first episode with similar characteristics, but only on the left side and that two years later he had a second picture, but this time, the problem occurred in the right ear, having been treated in both cases as acute cellulitis.

A biopsy of the left auricle nodule was requested and sent for histopathological study with a presumptive diagnosis of cellulitis vs. relapsing polychondritis.

The histopathological study reported normal appearance of the epidermis. The dermis shows a mild superficial perivascular inflammatory infiltrate with a predominance of lymphocytes and dilated capillaries (Fig. 5). The underlying cartilage shows adequate architecture with foci of eosinophilic degeneration towards the periphery and infiltrate close to the cartilage (Fig. 6). These histological changes are compatible with Relapsing Polychondritis.

With this diagnosis, laboratory tests were requested with the intention of initiating treatment with dapsone, which were found to be within normal limits and G6FDH within the usual limits.

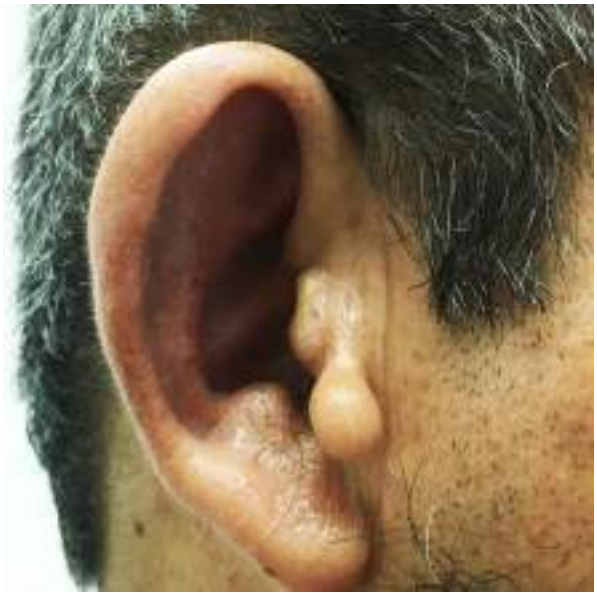


Figure 1. Erythema and inflammation of the right auricle. Respect of the lobe. Presence of pedunculated tumor arising in the anterior area of the tragus.

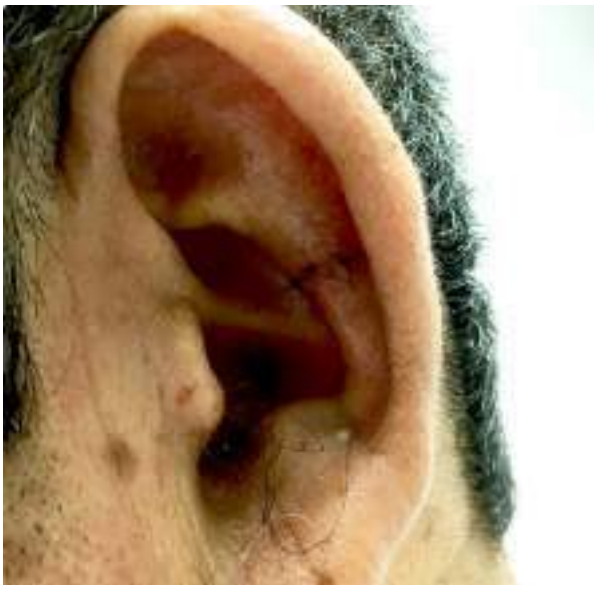
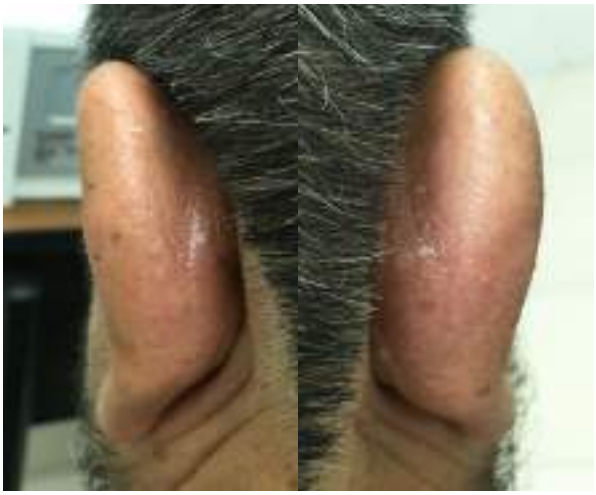


Figure 2. Erythema and inflammation of the left auricle. Respect of the lobe. The biopsy site is observed.



Figures 3 and 4. With the same disposition of the previous photos, a posterior view shows a notorious edema with a great increase in volume of both ears.

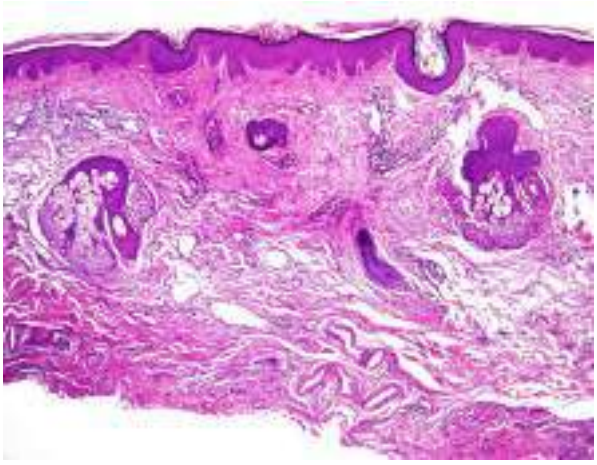


Figure 5. Dermis with superficial perivascular infiltrate with dilated capillaries.

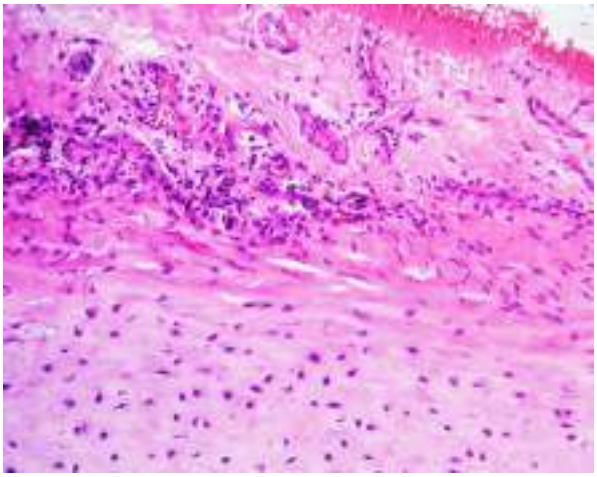


Figure 6. Underlying cartilage with foci of eosinophilic degeneration towards the periphery and infiltrate close to the cartilage.

Treatment was started with dapsone at a dose of 50 mg (1/2 tablet daily) for 7 days and then 100 mg/d (1 tablet) after lunch, 1 mg of folic acid was added daily orally and the patient was scheduled for evaluation in 40 days.

The patient did not return on the indicated date, but 5 months later, indicating that he took the medication for a month and a half and that he had practically total improvement with no recurrence so far. During the physical examination, only a slight increase in the size of the right ear and absence of inflammation was observed. The patient was prescribed topical application of pimecrolimus 1% at night for one week and then BID for two months. The patient has not returned for follow-up.

DISCUSSION

Of Relapsing Polychondritis

The etiology of RP remains obscure, multiple theories have been invoked, from a genetic predisposition demonstrated in the Ir region of the human histocompatibility antigen (HLA) class II[6] or, in another case by association with HLA/DR4/DQ8 in antigen presenting cells and CD4T helper positive lymphocytes at sites of injury.[7-8] Finally, several reports suggest that events such as mechanical, chemical, infectious and cancer aggressions, directed to cartilaginous structures, can provoke the appearance of RP in genetically susceptible patients.[8] Borgia et al. in their review article also emphasize theories related to circulating antibodies against collagens II, IX and XI in patients with RP, suggesting that cartilage-specific autoimmunity may play a crucial role in the pathogenesis of RP and, considering that type II collagen represents 95% of the total collagen content of cartilage, it is understandable that it may represent a very important primary target of autoimmunity and, in fact, antibodies against CII have been detected in one third of RP patients. Other accepted target antigens include; matrilin -1 and cartilage oligomeric matrix proteins, variations in their levels have been detected during episodes of RP, probably reflecting increased release from damaged cartilage.[9]

Clinically, the most characteristic and common finding is atrial involvement, but as noted above, other body sites may be involved. Atrial involvement is seen in approximately 90% of cases and the attack is strictly restricted to the cartilaginous fragments of the atrium with total respect to the lobes.

Patients usually seek medical attention for changes in ear color, tenderness and/or pain. The episodes present a pattern with relapses and remissions and can leave as sequelae deformity of the auricle, being of note that in a third of patients with RP there is ear involvement with hearing impairment, tinnitus and vertigo.[10]

There are no pathognomonic signs that lead to the diagnosis of RP, in fact it is made by the sum of several facts that have been gathered by several authors to elaborate several diagnostic criteria such as those of Mc Adam et al. in 1976,[11] Damiani and Levine in 1979[12] and Michet et al. in 1986.[13] The first one requests 3 of 6 clinical facts for the diagnosis and histopathology is not indispensable, on the other hand Damiani includes it and requests then the sum of one of the 6 clinical criteria of Mc Adams plus the positivity of histopathology and added to this the therapeutic response to DDS or corticotherapy to confirm the diagnosis of RP.[10]

Summarizing then, Mc Adam, in 1976 issues his criteria[11] and specifies that:

The diagnosis of relapsing polychondritis is made in patients presenting three or more of the following clinical features:

1. Recurrent bilateral atrial chondritis
2. Nasal chondritis
3. Non-erosive inflammatory polyarthritis
4. Ocular inflammation (keratitis, uveitis, scleritis and/or episcleritis).
5. Laryngo-tracheal-bronchial inflammation.
6. Hearing impairment (sensorineural loss, vertigo and/or tinnitus).

In 1979 these criteria were revised by Damiani and Levine[12-14] who redefined diagnostic positivity as follows:

1. The patient demonstrates three or more of McAdam's criteria. Histological confirmation is not necessary.
2. The patient demonstrates one or more of the McAdams criteria with histologic confirmation.
3. Patient has chondritis in at least two separate anatomic locations that respond to corticosteroid treatment.

It is interesting that RP has cutaneous manifestations in 14 to 37% of the cases and that several authors have pointed out,[14-15-16-17] among them, purpura, papules, nodules resembling erythema nodosum, aphthosis, urticarial eruption, ear ulcers, livedo reticularis, erythema elevatum diutinum, pustulosis, superficial phlebitis, ulcers of the limbs, distal necrosis. It should be noted that cutaneous manifestations are sometimes the first manifestation of RP in 12% of cases.[18] It should not be forgotten that the picture also presents ocular, respiratory, musculoskeletal, hematological, cardiovascular, neurological and renal manifestations.[19]

The histopathological study frequently shows cellular infiltrate of lymphocytes, neutrophils and plasma cells, more evident in the cartilage-skin interface, as well as the reduced number of chondrocytes in the areas of cartilage destruction. In general the cartilage biopsy is similar in all organs examined. There is loss of matrix mucopolysaccharides and the cartilage loses its basophilic staining with hematoxylin and eosin. There is perichondrial inflammation involving mainly lymphocytes, but neutrophils may be predominant in early lesions.[20]

At least one third of patients with RP show associated diseases of various kinds[10] with the list of associated diseases being quite long and including autoimmune disorders such as SLE, systemic sclerosis, mixed connective tissue disease, Sjögren's syndrome, dermatomyositis, vasculitis.[21] An increasing number of cases of RP related to malignant diseases have been described, especially myelodysplastic syndrome and, although less frequently, solid tumors (bladder, breast, lung, colon, pancreas) or other hematologic neoplasms such as lymphoma. There are reports of association with palmoplantar pustulosis,[22] Sweet's syndrome,[23-24] erythema nodosum.[25]

Differential Diagnosis of RP

Chondritis or inflammation of the external ear can also occur with trauma or infection, however in the latter cause, the earlobe is usually attacked, while RP is always spared. If the auricular chondritis is bilateral, recurrent, resolves spontaneously and is associated with other characteristics of RP, the diagnosis of RP can be easily made, but if the process is unilateral, a Pseudomonas aeruginosa infection with necrotizing otitis externa should always be considered.[20]

Accessory Tragus

We have already indicated that in the right ear at the level of the tragus there was a tumor clinically compatible with AT. During the embryological stage, the auricles originate and develop from six mesenchymal proliferations on day 42 of gestation, which are located at the dorsal ends of the first and second branchial arch, surrounding the laryngeal cleft. During embryological development, the first gill arches grow ventrally towards the midline to form the mandible. The tragus, the only area of the pinna derived from the mandibular arch, develops from the dorsal portion. The TA is located along the migration of the first gill arches. Initially they are located in the lower part of the neck, when the lower jaw develops, they rise until they are located in the lateral areas of the head at the level of the eyes, to later fuse and give origin to the ears.[5]

From the first gill arch derive the tragus, the helix and the superior branch of the antehelix and from the second the concha, the antehelix and the antitragus are formed.

Generally it is presented as a solitary lesion, but it can be multiple,[26] uni or bilateral, being the most frequent presentation at the level of the preauricular region near the tragus and they can also have other locations such as the neck[27-28] along the sternocleidomastoid muscle. However, these patients should be examined since they can be associated with other malformations or with other syndromes (Goldenhar, Townes-Brocks, Treacher-Collins Wolf-Hirschhron, Delleman, Noonan, VACTERL association, etc).[26-29-30-31]

These lesions are generally classified according to location and presentation patterns. According to the location they have been divided into two groups, the first corresponds to lesions located on the face and the second to those located in the auricular subunit, and these two groups in turn are subdivided into other subgroups according to the area taken either from the face or mouth, or on the other hand according to the anatomical area involved in the pinna.

According to the presentation pattern they have been divided into (Fig. 7):

(a) Pedunculated which in turn are subdivided into:
- Spherical
- Ovoid
- Lobulated
- Nodular

b) Sessile
c) Areolated
d) Remnant
e) Depressed

Since it is a minor congenital malformation and of benign character, treatment is not really necessary, if for aesthetic reasons its elimination is desired, surgical excision must be complete to avoid complications especially when the anomaly is deep and always avoiding excisions with shaving method or similar.[28]

In our patient the lesion is located within the pedunculated forms and in the ovoid subtype.

TREATMENT

As far as TA is concerned we emphasize that it is not really necessary except for esthetic reasons.

As for RP, mild forms of the disease only require anti-inflammatory drugs or oral corticosteroids in low doses. The more severe or refractory forms are treated with dapsone or prednisone in higher doses, there are reports of very severe cases in which immunosuppressive therapy (MTX, azathioprine, mycophenolate mofetil) has been used and even publications of cases treated with biologic therapy (infliximab etanercept, tozilizumab, rituximab).[4-33]

Patterns according to the form of presentation (taken from Zeumer M. et al. in[32])

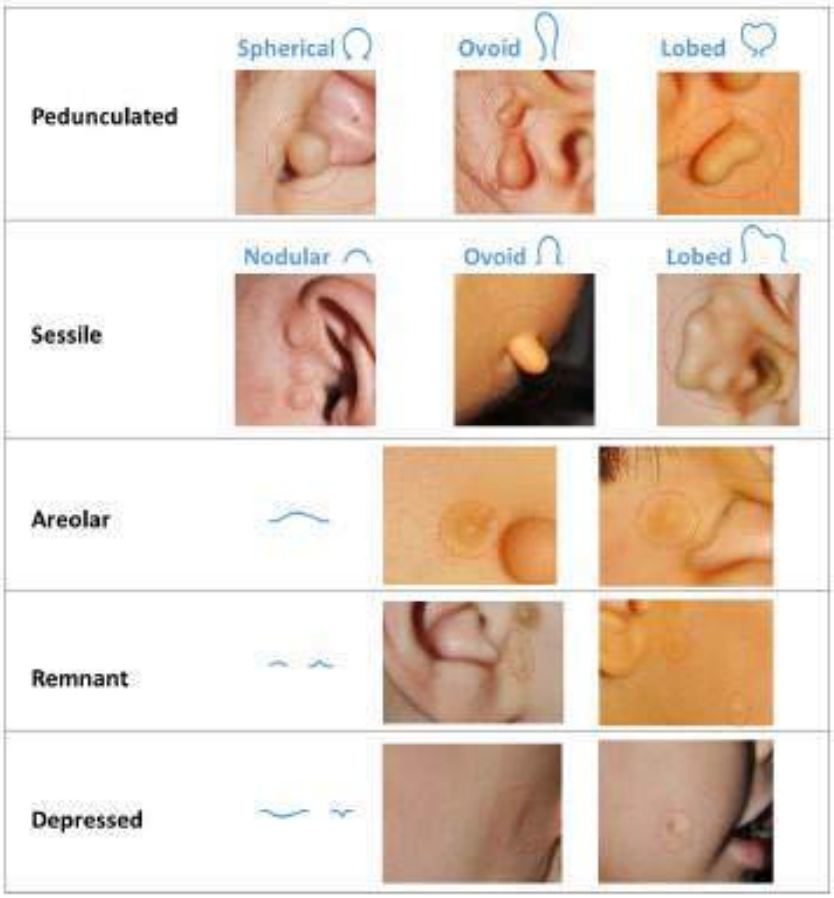


Figure 7. Taken from (32). The patterns of presentation of AT can be seen visually. Our case would correspond to the ovoid pedunculated form.

CONCLUSION

We have presented a case of association of AT and RP, in the literature to our reach we have not found reports in this regard. The diagnosis of RP was made based on the clinical aspect, episodic evolution, initially unilateral, changing the side of the affected pinna in each outbreak to finally become bilateral, the respect of the earlobes, the symptomatology according to the picture and the histopathological study that supports the diagnosis, thus fulfilling the criteria modified by Damiani, associated to a good therapeutic response with dapsone. The presentation of the two pictures in the same location, i.e. in the pinna, is very infrequent, which supports our interest in this communication.

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