Cutaneous Angiomyolipoma of the Ear: A Rare Diagnostic Challenge

Abstract

Cutaneous angiomyolipoma is an extremally rare mesenchymal tumor, distinct from its renal counterpart. We describe two patients with cutaneous angiomyolipoma of the earlobe, all of which were clinically misdiagnosed and mistreated as epidermoid cysts, to highlight the need to consider a broad spectrum of differential diagnoses when evaluating any nodular or cystic tumor of the ear. We also provide a comprehensive literature review about this cutaneous entity.

Keywords: *Angiomyolipoma, diagnosis, ear*

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Introduction

Dermatologists, surgeons, and general practitioners often encounter a wide spectrum of ear tumors. The clinical diagnosis becomes difficult due to similar morphologic appearances of some of these tumors. We describe two patients with cutaneous angiomyolipoma of the earlobe, all of which were clinically misdiagnosed and mistreated as epidermoid cysts, to highlight the need to consider a broad spectrum of differential diagnoses when evaluating any nodular or cystic tumor of the ear. We also performed a literature review for the cutaneous angiomyolipoma of the ear.

Case Report

Two adult patients presented with a history of slow-growing asymptomatic tumors in the earlobe. General practitioners diagnosed them with epidermoid cysts, and because of unsuccessful surgical drainage and antibiotic therapy, they were referred to our unit. Physical examination revealed nontender soft tumors in the earlobe on palpation [Figure 1a-d]. No epidermal punctum was present on the surface of the lesions. No sign suggestive of shagreen patch or hypopigmented macule was evidenced. None of the patients had any known personal or familial history of tuberous sclerosis. Clinically, we suspected that the lesions were lipoma and angiomyolipoma in the first and second patient, respectively. They underwent complete surgical excision [Figure 2a-c]. Histopathologic examinations were compatible with cutaneous angiomyolipoma [Figure 3a and b]. They had no recurrence or development of other similar lesions during follow-up [Table 1].

Discussion

Angiomyolipomas, benign tumors composed of blood vessels, smooth muscle bundles, and adipose tissue in varying proportions, are most commonly found in the kidneys of patients with tuberous sclerosis, whereas extra renal angiomyolipoma has been rarely described.[1] Cutaneous angiomyolipomas, an extremely rare form of presentation, have been reported in the toe, anterior abdominal wall, elbow, forehead, nose, chin, and ear. According to a comprehensive English literature review (Medline, Embase, SciELO, and LILACS databases) performed by two independent authors (disagreements resolved by consensus) in March 2019, by searching the databases using the terms "angiomyolipoma" and "angiolipoleiomyoma" in combination with the terms "ear", "auricle", "auricular", "earlobe", "lobule" and other related terms without date restrictions, 17 patients (including our two patients) with cutaneous angiomyolipoma of the ear have been described to date [Table 1].[1-13] All cutaneous angiomyolipomas of the ear occurred in adults aged 24–67 years.[1-13] Signs of tuberous sclerosis were absent in all the reviewed patients.[1-13] Cutaneous angiomyolipomas commonly presented as

How to cite this article: Araujo KM, Denadai R, Denadai R. Cutaneous angiomyolipoma of the ear: A rare diagnostic challenge. Indian Dermatol Online J 2020;11:226-8.

Received: August, 2019. **Accepted:** September, 2019. **Published:** March, 2020.

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Access this article online

Website: www.idoj.in

DOI: 10.4103/idoj.IDOJ_296_19

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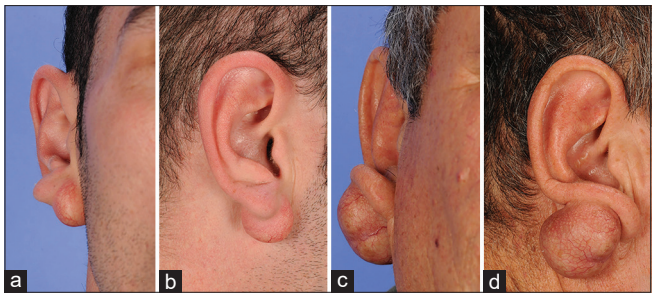

Figure 1: Clinical photographs showing earlobe tumors (Patients 1 [a and b] and 2 [c and d], Table 1)

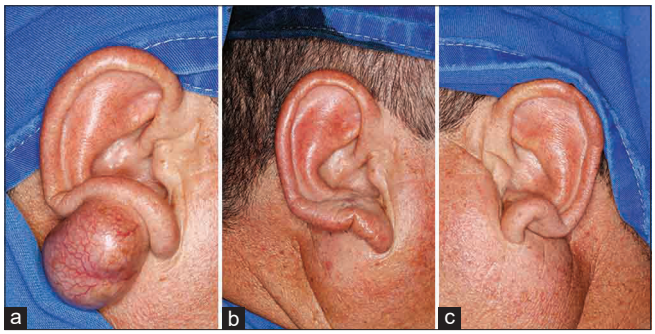

Figure 2: (a) Preoperative and (b) immediate postoperative views of surgical excision of the earlobe tumor under local anesthesia. (c) Non-affected ear (Patient 2, Table 1)

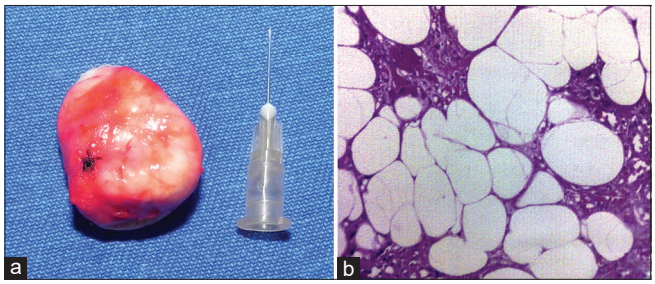

Figure 3: Histologic analysis of the (a) surgical specimen shows cutaneous angiomyolipoma (well-capsulated soft mass with clear boundaries). (b) An admixture of mature adipocytes, blood vessels, and scattered bundles of smooth muscles (Hematoxylin and eosin stain; ×400). No cytologic atypia or mitotic figures or HMB-45 immunoreactivity were noted, and there were no areas of necrosis or hemorrhage

solitary painless nodules in the helix or earlobe, giving the clinical impression of a wide spectrum of cystic or nodular lesions.[1-13] Most reported cutaneous angiomyolipomas of the ear were misdiagnosed as other cystic or nodular mimickers, including epidermal cysts, mucoid cysts, lipomas, or vascular tumors.[1-13] We report two patients with cutaneous angiomyolipomas misdiagnosed as epidermoid cysts by general practioners, leading to undertreatment. In our first patient, our clinical hypothesis was lipoma, and in the second patient, angiomyolipoma, given the similarity with the previous lesions. Similarly, a previous report[7] describes two cutaneous angiomyolipomas of the ear, for which the first clinical diagnosis was an angioma and in the second, angiomyolipoma, due to the similarity between the ear lesions.

These misdiagnoses were probably due to the rarity of the condition, the lack of awareness of the physicians, and insufficient clinical information, such as absence of detailed personal medical records and physical examination (e.g., an epidermal punctum has been described as a hallmark of clinical diagnosis of epidermoid cysts). The limited focus on cutaneous angiomyolipoma in standard textbooks compared to other cutaneous lesions may partially explain the lack of knowledge. Furthermore, with the rapid advancement and available imaging techniques and laboratory tests, there is a tendency among some physicians to rely more on technological reports for diagnosis rather than on the history, physical findings, and clinical judgment. This is relevant in different clinical settings, including the patients reported here, because there were no specific clinical, laboratory, and/or radiologic diagnostic criteria that aided in diagnosing cutaneous angiomyolipoma.[1-13] Although rare, our literature review suggests that physicians should be aware of the possibility of a cutaneous angiomyolipoma mimicking a broad spectrum of common and rare ear lesions and should consider it during the differential diagnoses when evaluating any patient with nodules, cysts, or cyst-like masses of the ear.

Complete surgical excision of cutaneous angiomyolipoma is both diagnostic and therapeutic. On histopathological examination, the diagnoses of all the reviewed cutaneous angiomyolipomas of the ear were confirmed on the basis of the classical triphasic traditional criteria, namely, blood vessels, smooth muscle cells, and adipose tissue. In addition, unlike renal angiomyolipomas, which are positive for HMB-45 immunochemistry stain, cutaneous angiomyolipoma shows no such reactivity.[1-13] As the reviewed cutaneous angiomyolipomas were well-circumscribed masses, they were effortlessly excised from the surroundings soft tissues. Most patients had no relapse during follow-up. Two relapses after surgical resections were described in one patient secondary to incomplete excision.[5] Therefore, complete surgical excision is curative and of paramount importance to prevent relapse.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Table 1: Characteristics of patients with cutaneous angiomyolipoma of the ear Published in English Literature[1-13]

Author, country, Year (reference)	Gender, age (years)	Tuberous sclerosis	Anatomical location, side	Size (cm)	Duration	Clinical Diagnosis	Treatment	Follow-up	Relapse
Argenyi *et al*., USA, 1991[1]	M, 67	No	Helix, R	1	40 years	Epidermal cyst	Surgical excision	5 years	No
Mehregan *et al*., USA, 1992[2]	M, 49	No	Helix, R	-	-	Epidermal cyst	Surgical excision	-	No
Lee *et al*., Korea, 1996[3]	M, 32	No	Earlobe, L	1.2×1.5	5 years	Lipoma, Epidermal cyst	Surgical excision	-	No
Val-Bernal and Mira, Spain, 1996[4]	M, 49	No	Earlobe, R	2	5 years	Lipoma, cyst, vascular tumor	Surgical excision	-	No
Büyükbabani *et al*., Turkey, 1998[5]	M, 38	No	Retroauricular area, R	2.5	10 years	-	Surgical excision	-	Yes†
Beer, Australia, 2005[6]	M, 43	No	Ear, L	0.4	6 months	-	Surgical excision	23 Months (mean)	No
	F, 44	No	Helix, L	0.5	3 months	Cyst			No
Sánchez-Estella *et al*., Spain, 2009[7]	F, 58	No	Postauricular region, L	1.5	5 years	Angioma	Surgical excision	26 months	No
	F, 52	No	Postauricular region, L	1	2 years	Angiomyolipoma	Surgical excision	5 months	No
Shin *et al*., Korea, 2009[8]	F, 26	No	Helix, L	1×0.9	Several years	Mucoid cyst	Surgical excision	3 months	No
Mikoshiba *et al*., Japan, 2012[9]	M, 37	No	Earlobe, R	1.7×1.6	Several years	-	Biopsy	-	-
Hanson *et al*., USA, 2012[10]	M, 24	No	Ear, R	-	-	-	-	-	-
Yaşar *et al*., Turkey, 2014[11]	M, 67	No	Earlobe, R	2×2	10 years	-	Surgical excision	2 years	No
Tchernev *et al*., Bulgaria, 2014[12]	F, 66	No	Helix Ear, R	-	Several years	-	Surgical excision	4 weeks	-
Mannan *et al*., USA, 2019[13]	M, 36	No	Ear, R	1.8×1.5	-	-	Surgical excision	-	-
Index case: Araujo *et al*., Brazil, 2019	M, 32*	No	Earlobe, R	1.3×1	4 years	Lipoma	Surgical excision	44 months	No
	M, 52*	No		2.6×2.2	6 years	Angiomyolipoma		28 months	No

M = Male; F = Female; R = Right; L = Left; - = unknown/not available; *Patients 1 [Figure 1a] [Figure 1 b] and 2 [Figure 1 c] [Figure 1 d] in the sequence; †Two previous relapses following surgical excisions

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