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Epithelioid Hemangioma In Jugal Location Mimicking An Arterial Malformation: A Case Report.

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Summary:
The clinical similarity and the panoply of names that presents the vascular abnormalities can lead us to see how it's still difficult to differentiate them. Epithelioid Hemangioma is one of them. This tumor can arise in subcutis, soft tissue and bone, and may resemble to other lesions including malignant.
Here, we report a case an EH involving the jugal region from his diagnosis to his treatment and we review the literature about soft tissues localization.

Keywords: *epithelioid hemangioma, differential diagnosis, Jugal, Angio scanner, Embolization, Surgery*

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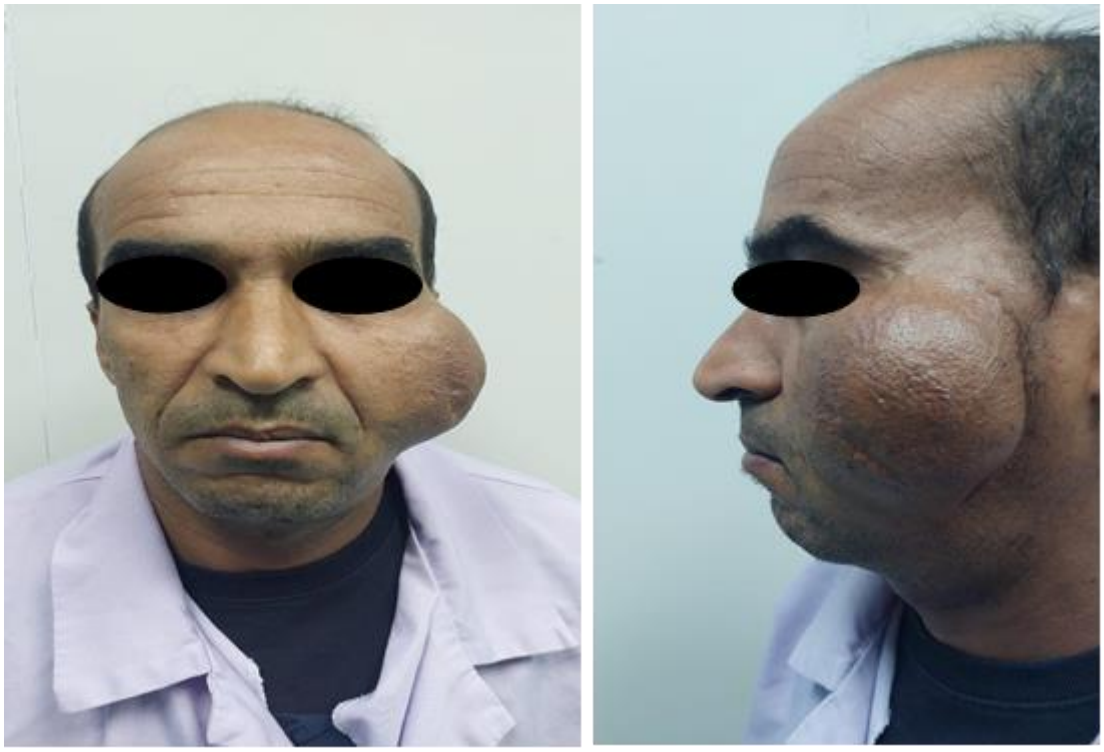
I. Introduction

Described for the first time by Wells and Whimster in 1969 under the name of Epithelioid Hemangioma (EH). The EH is vascular neoplasm characterized by the presence well-formed blood vessels lined by plump, epithelioid (histiocytoid) endothelial cells, with abundant eosinophilic cytoplasm and a variable eosinophilic infiltrate. (2) Even if this tumor is benign, his clinic presentation is unspecified and can mime other vascular tumors. His diagnostic of certitude is histological and the main treatment surgery.

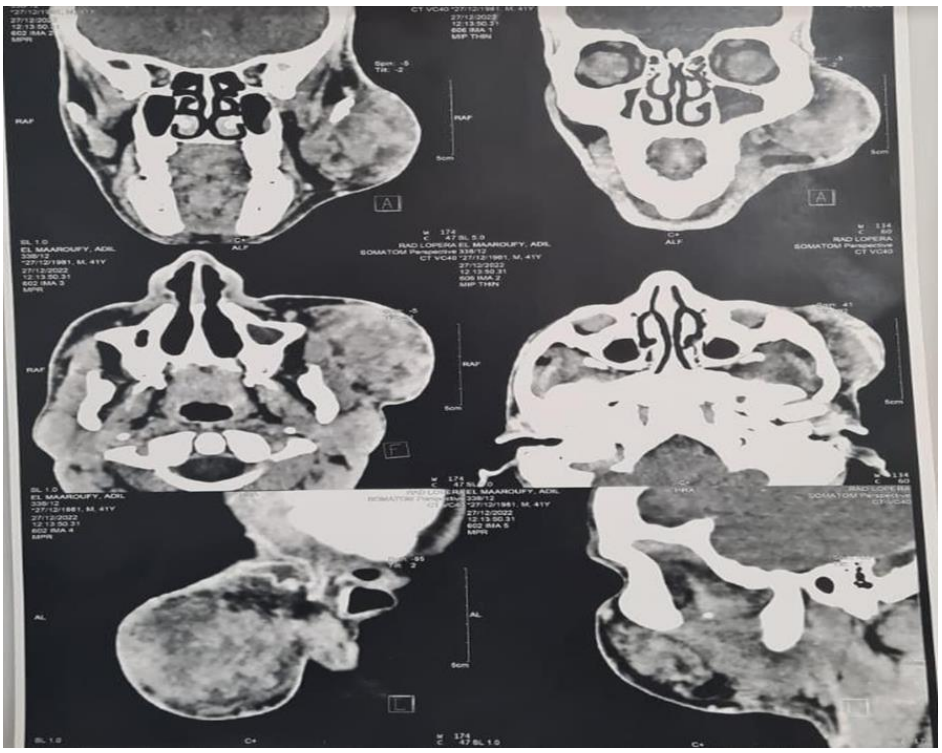
II. Materials And Methods:

We present here the case of a 43-year-old patient, who consulted us for a slow growing painless left jugal tumor that had been present for approximatively 20 years. The history revealed surgery for an undocumented left jugal mass at the age of 12 and reappear 10 years later. No other surgical or medical history was found. The patient was an active smoker, with 20 packs a year.On clinical examination, a roughly oval mass measuring approximately 8 to 9 cm (in his long axis) on the left cheek was found. It was firm, painless, renitent, throbbing, without thrill or pruritus, and adherent to the deep layers. The skin opposite was healthy, with an orange peel appearance associated with the scars of the previous surgery. There were no sensory disturbances or facial paralysis. (Fig. 1)



The rest of the clinical examination did not reveal any adenopathy in the head and neck region or any other pathology.

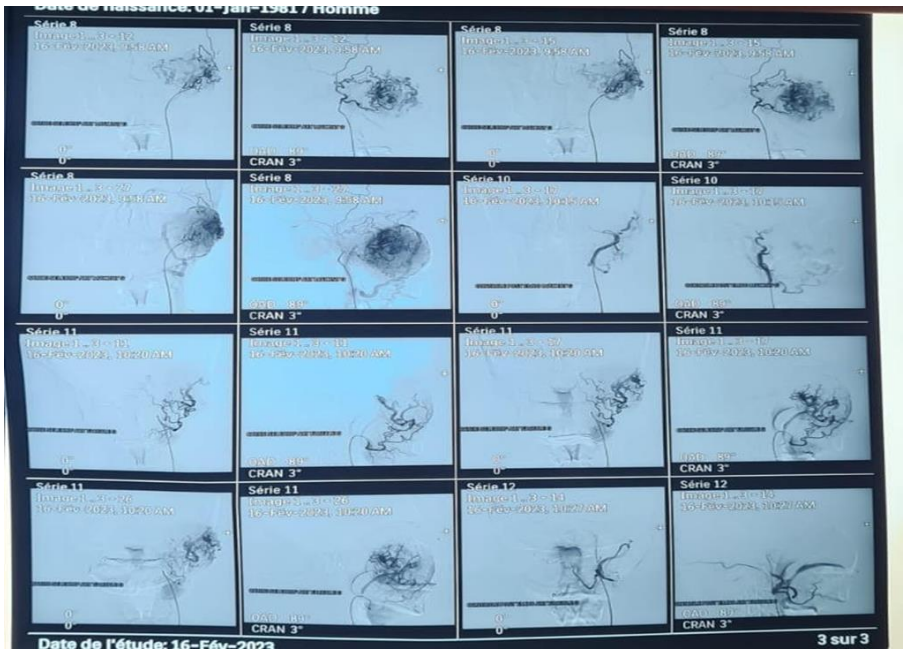
In the light of the clinical findings, an injected CT scan of the facial mass was performed to determine the relationship of the mass to adjacent structures and whether it was vascular in nature or not. This suggested a heterogenous subcutaneous vascular tumor encountering the masseter muscle. (Fig. 2)



In consultation with the interventional radiology team, an arteriogram was carried out and concluded that the patient had an arteriovenous malformation fed by branches of the left internal maxillary and facial arteries. Fig.3

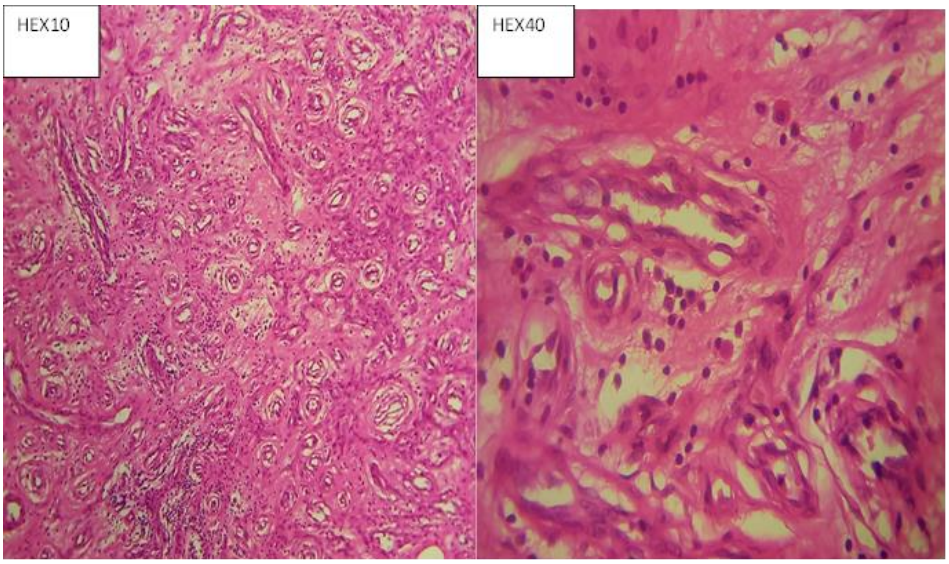
After a selective embolization of these two arteries with ***Curaspon®***, the patient was admitted to the operating theatre for surgical resection under general anesthesia, resuming the old incision.

The operation was performed without ***curare*** to facilitate the dissection of the facial nerve branches, which was the main obstacle to this surgery. The lesion was poorly defined and adhered to the skin tissue and deep muscles plane, with multiple sinuous vessels.

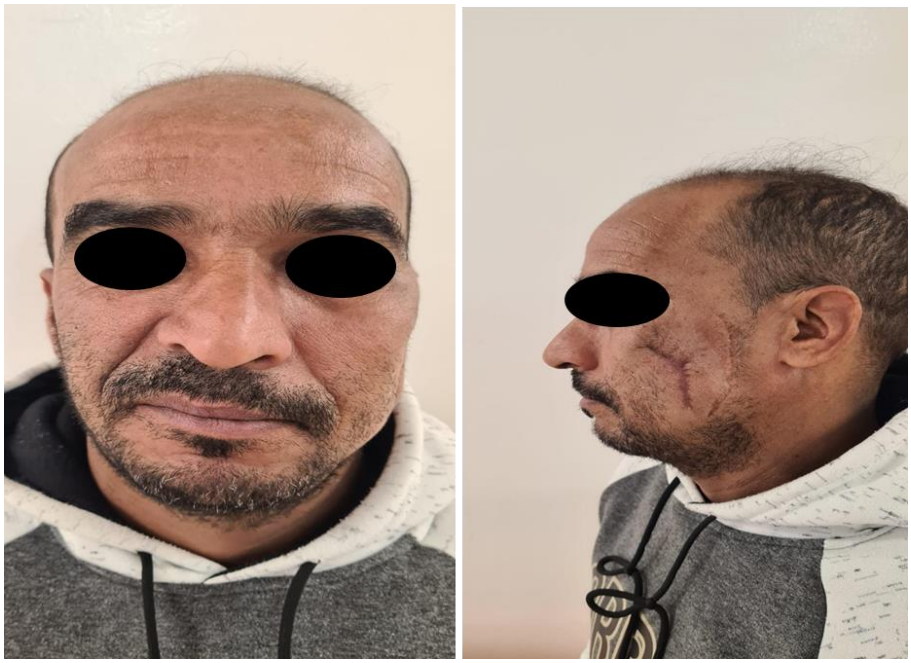


The anatomopathological study concluded to the diagnosis of epithelioid hemangioma.

Histologically, it was composed of lobular proliferation of small, capillary-sized vessels lined by plump and epithelioid endothelial cells with abundant eosinophilic or amphophilic cytoplasm that appear to project into the lumen. An abundant stromal chronic inflammation with numerous eosinophils, lymphocytes, mast cells, and plasma cells was observed. (Fig.4)



At 5 months postoperatively, the results are satisfactory despite upper labial paresis for which he underwent for reeducation and there's no early recurrence. (Fig 5)



III. Discussion:

Described for the first time as Epithelioid Hemangioma by Wells and Whimster in 1969 under the name of Epithelioid Hemangioma (EH). In literature, this pathology is known under other terms like pseudo pyogenic granuloma, atypical pyogenic granuloma, inflammatory angiomatous nodule (2, 6). The term subcutaneous angioblastic lymphoid hyperplasia with eosinophilia and histiocytoid hemangioma is not recommended by the recent 5th edition of the WHO classification of soft tissue and bone tumors to define EH. It is a vascular anomaly recognized by the World Health Organization and International Association for the Study of Vascular Abnormalities (ISSVA) as a benign tumor. (1 4)

EH can affect subcutis, bones and soft tissues including organs like Penis, heart. (6)

The cause of HE remains unclear. Some authors believe that HE is a neoplastic lesion of endothelial cellular origin while others consider as a reactive phenomenon secondary to a traumatic attempt of an artery or a vein, or a hormonal imbalance. (2, 5,8)

This rare tumor can be seen at any age with the predominance between the third and six decades. Concerning the gender it's controversial and if we look more closely, we can see that depending on the tissue affected, the sex ratio differs. Thus, in subcutaneous locations affect more men when those affecting bone and soft tissue have a women predilection. (3,6,8,10)

EH are described to be more frequent in head and neck region, predominantly in the periauricular area. But some authors support a predominance of subcutaneous involvement of the limbs. (2,3,6,9)

The clinical presentation is variable. Most commonly, the lesions affecting the subcutis appear as a single or multiple lesions in the form of papules, subcutaneous angiomatous nodules or erythematous plaques that are painless in general, sometimes hemorrhagic, or pruritic, and always progressive. They can also show fluttering like in our case. (6,9)

HE must be differentiated from other pathologies with similar presentations, such as mesenchymal tumors of the vessels, nerves or muscles of the head and neck like (neurilemmoma, leiomyoma, hemangioma and fibrosarcoma); Kimura disease; epithelioid hemangioendothelioma; epithelioid angiosarcoma; BANGAL intravascular papillary endothelial hyperplasia and vascular malformations. (13)

As clinically, there's no specific radiological images most of which is reported is in the form of case reports because of the lack on the imaging features of epithelioid hemangiomas. Preoperatively, CT and MRI are often used to evaluate HE, as well as angiography with possible embolization. (10,12)

Histological evaluation is therefore essential for the diagnosis of EH.

Histologically, HE in subcutis appears as a cluster of endothelial cells protruding into the lumen; cells have abundant, eosinophilic, hyaline cytoplasm and scalloped margins. Infiltration of immature inflammatory cells is also evident. Lymphocytes and eosinophils surround and can infiltrate obvious blood vessels. A lymphoid tissue may form reactive germinal centers, particularly at periphery of lesion. Mitotic figures are uncommon. There is no necrosis and no nuclear pleomorphism. (15)

Endothelial cells are positive for CD34, CD31, ERG, FLI1, and factor VIII-related antigen (9).

These morphological findings lead to the discussion of Epithelioid hemangioendothelioma. However, Epithelioid hemangioendothelioma is composed of Chains and cords of epithelioid cells distributed in a myxo hyaline stroma. The Cells contain intracytoplasmic lumina without well-formed vessels or abundant stromal eosinophils, plasma cells, and lymphocytes. (16,17)

The main treatment for HE is surgical excision; recurrence is attributable to insufficient surgical margins but also to early appearance of the disease in childhood like for our patient. (5,7,13 ,11)

treatments are available when anatomical considerations make complete excision impossible, or the lesion is diffuse, or recurrence develops. The medicines suggested are Oral propanol and high-dose steroids. Also have been tested cryotherapy and chemotherapy. Some others proposed the use of different types of lasers with variant results. (6,11)

Cytotoxic drugs such as bleomycin are useful in cases of steroid resistance or intolerable side effects. (14)

IV. Conclusion:

This plethora of terminology, in addition to the poor understanding of the pathogenesis, partly explains the confusion surrounding this vascular-inflammatory lesion. HE should be considered in the differential diagnosis of patients presenting with pulsatile masses in the oral and maxillofacial regions.

The mode of presentation of HE varies; the HE can imitate several other entities.

Histological studies are essential, and more research will help for a better understanding of this pathology.

Conflicts Of Interest:

The authors declare no conflict of interest.

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