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Epithelioid haemangioendothelioma arising in the nasal cavity

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Abstract

We report here the case of an epithelioid haemangioendothelioma (EHE) arising in the nasal cavity which is, to the best of our knowledge, the first ever described example in the world literature in that particular site.

The patient is a 23-year-old male who presented with repeated episodes of epistaxis from the nasal cavity and with a 1.5 cm reddish, polypoid, smooth, spontaneously bleeding nodule in the right middle meatus. This lesion was histologically diagnosed as epithelioid haemangioendothelioma. Immunohistochemically the neoplasm displayed striking positivity for CD31, CD34 and vimentin. A surgical approach was performed by 'facial degloving', removing the right inferior turbinate, the anterior two-thirds of the middle turbinate and the medial wall of the ethmoid bone. After 12 months follow-up the patient is disease-free, without any local or distant recurrence.

Key words: Haemangioendothelioma, Epitheloid; Nasal Cavity

Introduction

The term 'epithelioid haemangioendothelioma' (EHE) was first applied by Weiss and Enzinger in 1982[1] to define a special entity among vascular tumours. It now belongs to vascular tumours with an endothelial component, and it is considered a borderline lesion, between the fully benign entities, such as epithelioid haemangioma, and the malignant ones, such as epithelioid haemangiosarcoma.[2]

EHE is not a common lesion and its clinical course is unpredictable. Sometimes local recurrence and distant metastasis have been described. EHE can arise in different sites, such as most commonly, the skin, liver, lung, bone and (rarely) the oral cavity, gastrointestinal tract, peritoneum, lymph nodes, meninges, mediastinum, pleura, heart, spleen, thyroid and parotid gland.[3–6] It has never been reported in the nasal cavity. We describe the case of a vascular tumour in the nasal cavity of a 23-year-old male, classified and diagnosed as EHE by histological immunohistochemical criteria.

We have not been able to find a similar case in the literature.

Case report

A 23-year-old male was evaluated at the Otolaryngology Department – University of Rome 'Tor Vergata' in July 2000 for repeated episodes of epistaxis from the nasal cavity, associated with important sideropenic anaemia (Hb 7 mg/dl). He had already been treated elsewhere by nasal septum varices electrocoagulation, without any reliable benefit.

A fibro-rhino-endoscopy was performed, showing a small, reddish, polypoid, smooth, 1.5 cm spontaneously bleeding lesion, located *p + 19Xin in the right middle meatus. Neck lesions were not present. A computed tomography (CT) scan, showed the presence of a 1.5×1.5 cm mass in the middle meatum, not invading adjacent osseous structures (Figure 1).

A surgical approach was performed by 'facial degloving', removing the right inferior turbinate and the anterior two-thirds of the middle turbinate and the medial wall of the ethmoid bone.

Microscopically, the lesion showed the common spectrum of pathological features of EHE. The lesion had epithelioid and histiocytoid components, creating many neoformed spaces containing red blood cells, with nuclear vacuolization and vesiculation (Figure 2(a)). Mitotic figures were rare and mild nuclear atypia was seen (Figure 2(b)). Neoplastic cells were sparse in fibrous connective tissue. Immunohistochemical study, performed according to the method of Hsu *et al.*,[7] confirmed the endothelial nature of the neoplasm, with a striking positivity for CD31, CD34 and vimentin (Figure 3). Immunostains for histiocytic, epithelial cells and S100 were negative. The above findings were in keeping with the diagnosis of EHE.

After a 12-month follow-up period the patient is disease-free, without any local or distant recurrence.

Discussion

The term EHE has been used over years to describe both benign and malignant vascular tumours composed of endothelial cells and it lacks specificity. It was first described as a specific entity by Weiss and Enzinger in 1982,[1] showing, as peculiar features, endothelial cells with epithelioid and histiocytoid morphology. The cytoplasm of the proliferating elements is generally abundant, often vacuolated, with round, vesicular and indented nuclei.

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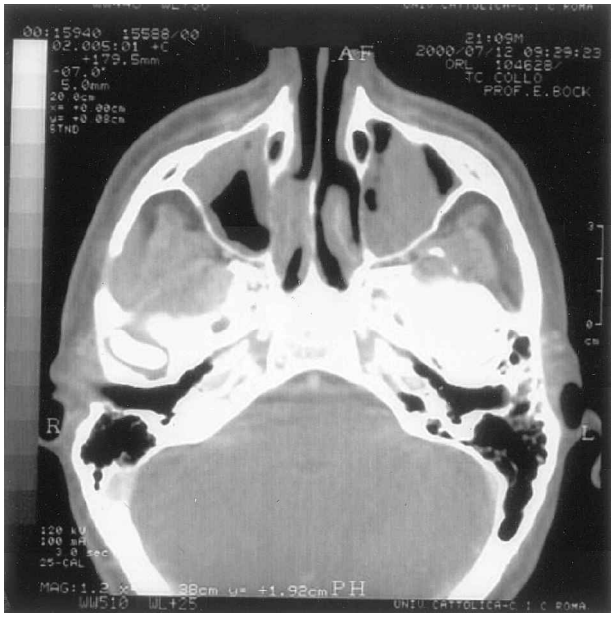
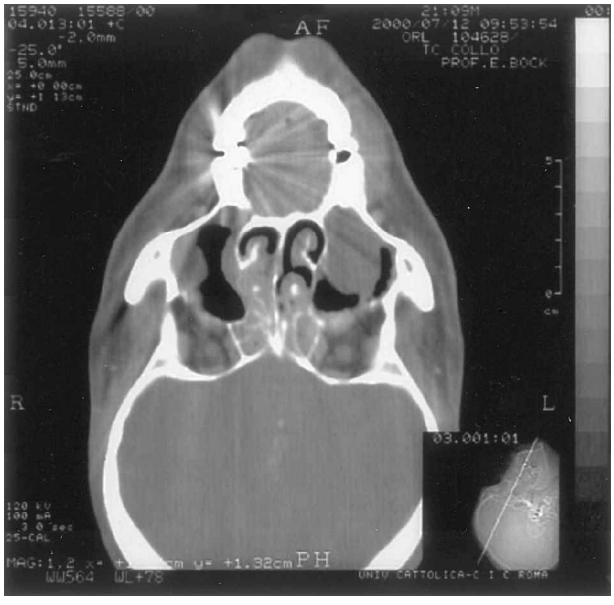


(a)



(b)

FIG. 1

Computed tomography scan of the nasal cavity and maxillary sinuses. (a) axial view showing bilateral chronic inflammatory thickening of the maxillary sinuses and a protruding mass in the right middle meatus. (b) In the coronal view the tumour is fully demonstrated. Chronic inflammatory changes of the maxillary and ethmoid sinuses are seen. Bone erosion is not evident.

Tumour cells expand the vessels, usually preserving their architecture as they extend centrifugally from the lumen into soft tissue. These cells form vascular lumina, commonly located intracellularly, producing cytoplasm vacuolization. The presence of erythrocytes and blood in vacuoles confirms the endothelial nature of these cells.[8] An inflammatory infiltrate can be present.[1,8,9]

EHE is not a malignant tumour, but it has an unpredictable clinical behaviour. In about 25 per cent of the described cases, it contains areas with significant atypia, mitotic activity (>1 mitosis per 10 high power field), spindling of cells and necrosis. It has been

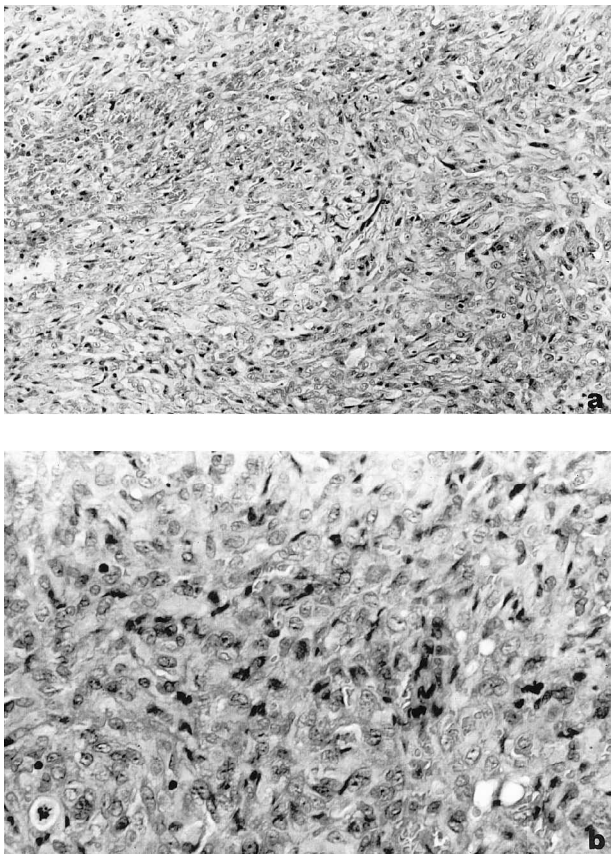


FIG. 2

Photomicrograph of EHE. (a) The tumour appears to be composed of solid sheets of plump, epithelioid cells with the presence of blood filled vascular spaces (H & E; ×250). (b) High power view of the tumour showing the epithelioid appearance of the proliferating endothelial cells (H & E; ×400).

suggested[6,10] that metastasis occurring in such cases has its origin in these cellular subpopulations. Immunohistochemical positivity for vascular markers such as CD31, CD34 and F VIII confirms the diagnosis. Rare focal positivity for cytokeratin, complicating differential diagnosis, may reflect the neoplastic cells' primitive nature. Moreover, a differential diagnosis with metastatic carcinoma must be considered, but nuclear pleomorphism, mitotic activity and immunohistochemical negativity for vascular markers will rule it out.[1]

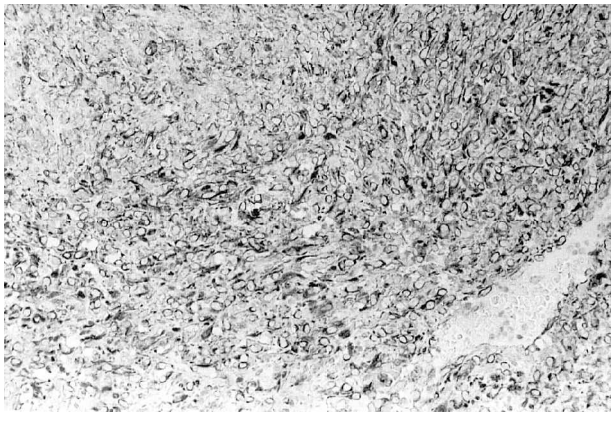


FIG. 3

Immunostaining for CD31 shows a strong and diffuse reaction in proliferating tumour cells (×250).

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Clinical behaviour and prognosis varies greatly, depending on the organ of origin and propensity for multifocality. In the largest series of described EHE,[6,8,11] 13 per cent of the cases recurred locally, 31 per cent had metastases (50 per cent in regional lymph nodes and 50 per cent in the lung) and 13 per cent died, within a four year period. The treatment of choice is wide local excision, which is curative in most cases. Radiotherapy alone is hardly ever effective due to slow growth of tumour cells.[9] For malignant variants, regional lymph node dissection is suggested.[8] Pre-operative diagnosis is not always easy, with the clinical picture showing just a solitary lesion.[2]

EHE has been described rarely in the head and neck namely in the thyroid,[5] larynx[12] and parotid gland.[3]

We describe here the first case of EHE of the world literature arising in the nasal cavity, diagnosed by histological, immunohistochemical and clinical data.

Radical local excision of the lesion with adequate clear margins was obtained, taking into account the risk of local recurrences described in this tumour type in other body areas. An external approach was then undertaken by 'facial degloving' to gain adequate exposure of the middle meatus area avoiding facial scars in a young patient.

Our patient is disease free at 12 months after initial diagnosis, but a careful follow-up has been recommended to avoid any local recurrence that, in this kind of neoplasm, is the most often feared complication.

In conclusion, the present case appears to be unique for its localization. The correct clinical approach and the appropriate surgical treatment remain to be defined due to the lack of previously reported cases. In view of the unpredictable clinical behaviour of EHE, it is important to keep in mind the existence of this type of tumour in the differential diagnosis of masses arising in the nasal cavity.

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