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Case Report

Epithelioid Hemangioendothelioma of Pterygomaxillary Fissure

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Abstract

Introduction

Epithelioid Hemangioendothelioma (EH) is an uncommon vascular neoplasm (endothelial origin) which can affect any type of tissue. It rarely occurs in head and neck region, and has unpredictable clinical behavior.

Objective

The aim is to report a case of EH of unusual location (pterygomaxillary fissure) and histological rarity, andto establish an accurate differential diagnosis of EH and Juvenile Nasoangiofibroma (JNA).

Case report

A 19 year-old male with a chief complaint of recurrent epistaxis and an obstruction of the left nasal cavity. The diagnosis of JNA was suggested and surgery with tumor complete resection was the election treatment. However, clinical diagnosis was not confirmed postoperaively, through Immunohistochemistry (IHC). Histopathological findings showed a diagnostic reporting of EH. Tumor had no recurrence within three years after surgery.

Discussion

Apart from the rarity of the condition, EH varies according to their location in asymptomatic or with few symptoms. The presence of violaceous lesions indicates a huge range of vascular tumors. Thus, IHC has assumed very important for diagnosis confirmation.

Conclusion

The localization of EH in perygomaxillary fissure is a rare condition withpoor prognosis. The treatment choice for EH was the complete ressection.

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Introduction

Epithelioid Hemangioendothelioma (EH) is a rare tumor of endothelial origin. It can develop in any body tissue, and the most common sites are soft tissues, liver and lungs [1,2]. EH tumor was first described, in 1975, in lung and was denominated "intravascular bronchioloalveolar tumor" [2]. Later, in 1982, [3] EH was reported in liver as a distinct entity with unpredictable malignant potential [2,3]. HE tumor has borderline malignancy and agressiveness [4]. Biological behavior of a EH is intermediate between hemangioma and conventional angiossarcoma. It has local recurrence in one third of tumors, and in a few cases occurs the process of metastasis [5]. EH growth is slow and progressive, and tumor regression can also be observed spontaneously in some cases [5]. The peculiarity of this tumor is the occurrence in veins of medium and large size which features well-defined vascular channels. Clinically, it presents areas of fibrosis, calcification, focal necrosis, hemorrhage and cystic degeneration. Histologically, it shows vascular channels with masses, sheets of spindle cells and often cuboidal, and sometimes with mitotic figures [6]. These histologic features may induce a false-positiveprognosisfor: hemangioma, angiosarcoma and other conventional vascular tumors [2]. Genetic or environmental factors are not correlated with EH tumor arising, however an association with the use of estrogens and steroid hormones have been described [2]. Resection treatment cures most of tumors, but even then 40% relapse and 20% present with metastasis [6]. There is noeffective therapeutic strategies for HE and it is a tumor of poor prognosis [7]. The aim of this paper is to report a case of HE due to histological rarity and unusual location, and to establish a differential diagnosis of EH.

Case Report

A 19 year-old male presented signs of left-sided nasal obstruction and intermittent epistaxis for approximately one year. Nasopharyngolaryngoscopic exam revealed in left nasal cavity anangiomatous lesion with bleeding to light touch. Contrast enhanced computed tomography (coronal and axial sections) demonstrated that tumor included on left side: pterygomaxillary fissure, nasal cavity, maxillary and sphenoid sinuse, infratemporal fossa and upper limit near greater wing of sphenoid bone in high density (Figure 1). To confirm diagnosis hypothesis, a carotid angiography demonstrated an intense signal in deep face spaces, exactly in distal branches of internal maxillary artery, which is characteristic of Juvenile Nasoangiofibroma (JNA). The only tomography characteristic not related to JNA was pterygomaxillary fissure non enlargement. The preoperative diagnosis was tumoral blush, suggestive of JNA. Treatment was tumor complete resection through facial translocation (Figure 2).

However, immunohistochemistry (CD34 marker) of tumor resection demonstrated that tumor was an EH. The presence of cellular atypia and mitotic frames are compatible with malignancy. There were no signs of neoplasia in bone tissue (Figure 3). Follow-up exams, quarterly periodical, were: nasopharyngolaryngoscopic and contrast-enhanced computed tomography, and there was no evidence of recurrence in three years.

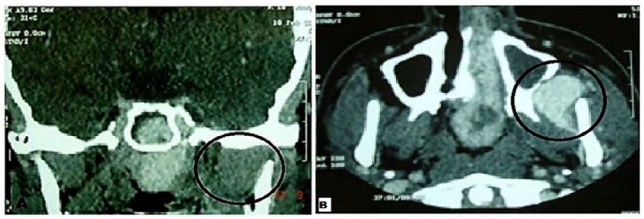

Figure 1A and 1B: Black circles in contrast-enhanced computed tomography (coronal and transverse sections, respectively) indicates HE localization in left side and structures envolved: pterygomaxillary fissure, nasal cavity, maxillary and sphenoid sinuses and upper limit near the greater wing of the sphenoid bone region.

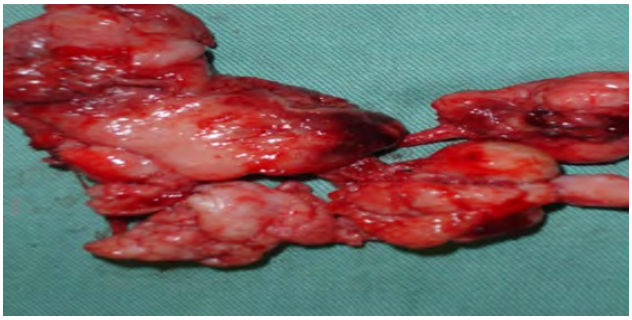

Figure 2: Surgical specimen.

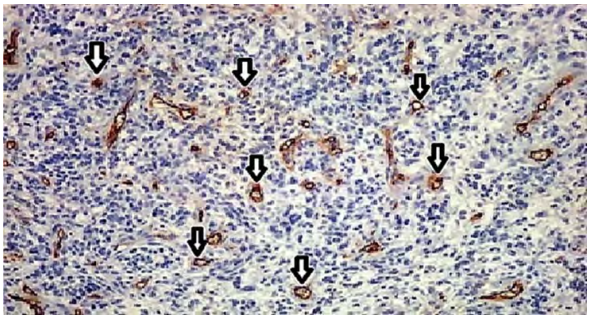

Figure 3: Immunohistochemistry with CD34 marker.

Black arrows indicate rich vascular network of tumor.

Discussion

Only two cases of HE in parapharyngeal region were described, until 2006: in submandibular space and in oral cavity [8]. A literature review and report of 160 articles published nine cases in head and neck region and only one case of HE in pterygomaxillary fissure [9]. In a recent review, mean age of the patients with HE was 28-year-old (4-to 76-year-old) and sex predilection was for females over males (2, 5:1) [5]. HE tumor may be asymptomatic or have few symptoms: temporal or retroauricular pain [10] and, spontaneous epistaxis. HE staining classically shows a violaceous lesion, with differential diagnosis of : epithelioid hemangioma, pyogenic granuloma and Kaposi's sarcoma. Our case report is a HE tumor with angiomatous lesion and some pathological findings like: atypical cells, cytoplasmic vacuoles, focal necrosis and squamous cells. These findings tend to form anastomoses, a characteristic of HE. According to the morphological criteria of stout: atypical endothelial cells outlining the vessel wall and tendency to form vascular lumen and anastomoses [11]. It is difficult to determine tumor grade in biopsies, since one single tumor can be classified in several different grades of malignancy [12]. Besides histopathological diagnosis, it is essential to perform specific immunohistochemical study-with frozen section-to confirm diagnosis from angiosarcoma [2,13]. Immunohistochemical analysis of vascular epithelial markers demonstrates positivity to form the factor VIII-related antigen, CD31, CD34 and vimentin [2,14,15]. Treatment recommended in literature for EH is the use of: antiangiogenic drugs, interferon alfa-2B, local embolization, radiation therapy, chemotherapy, and finally surgery (complete resection). Tumoral resection was necessary because of grade of malignancy. There is no consensus in the literature regarding the use of preoperative embolization, so this procedure was not used in our patient. Radiotherapy is not encouraged in the literature too, due to slow tumor growth [2], and it was not performed. A small number of patients with HE pathology in head and neck region reduces prognosis inferences. Cases of poor prognosis are located mainly in the liver and lung.

Conclusion

EH tumor in pterygomaxillary fissure is rare and of intermediate malignancy. Similarly findings in tomography and angiography exams demonstrate the difficulty in the differential diagnosis from: hemangioma, angiosarcoma and JNA. Complete resection is the elected operative treatment, since no recurrence was shown in three years.

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