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Case Report

Angiosarcoma in the Maxillary Sinus. How Frequent is it? A Case Report and Literature Review

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Abstract

Angiosarcoma is a rare of malignant and aggressive neoplasm characterized by the vascular endothelium growth that frequently occur in the elderly population. This entity can be presented in the head and neck region, especially in the superficial soft tissue of the scalp and face being the paranasal sinus an uncommon primary location for angiosarcomas accounting for less than 0.1% of all paranasal sinus malignancies. Epistaxis, nasal obstruction and nasal discharge are characteristic symptoms when this type of neoplasm is located in the paranasal sinuses. In this paper, we present a rare case of angiosarcoma in the maxillary sinus with an oral clinical manifestation as well as make a literature review of this unusual entity.

Keywords: Angiosarcoma; Paranasal Sinus; Vascular; Tumor

Introduction

Angiosarcomas are rare malignant tumors of the vascular endothelium, constituting 1.3% of soft tissue sarcomas. They are characterized by the formation of irregular vascular channels that are lined by one or more layers of atypical endothelial cells, usually with an immature appearance [1,2]. They may appear in any region where a blood vessel or lymphatic channel is found, which makes them common in cutaneous regions; however, their appearance on the head and neck is infrequent, with the scalp being the most common site of appearance [3,4]. These fast growing tumors can be clinically evidenced as an erythematous patch or an ill-defined purplish plaque, nodule or papule. The occurrence of this entity in the paranasal sinuses and specifically in the maxillary sinus is very rare, where its most relevant clinical characteristic is epistaxis, due to the vascularity of the tumor [5].

The objective of this study is to present a rare case of an angiosarcoma in the maxillary sinus with an oral clinical manifestation as well as make a literature review of this unusual entity.

Case Report

A 65-year-old male patient who attended the Oral and Maxillofacial Surgery Unit of Coromoto Hospital, Maracaibo - Venezuela, due to headache, nasal obstruction, epistaxis and left side gingivorrhagia, with 4 months of evolution without receiving treatment. The patient did not refer any relevant medical history; however, he reported 40 continuous years of work in the oil industry, specifically in the drilling area. On clinical examination, an increase in volume was observed in the middle third of the left side of the face, indurated on palpation, without signs of inflammation (Figure 1). Intraorally, it presented an oval, irregular increase in volume in the left palatal region, indurated on palpation, covered by

intact mucosa (Figure 2). A computed tomography was obtained and the total occupation of the left maxillary sinus was observed, with erosion of its walls including the ipsilateral alveolar recess as well as involvement of the soft tissue component, with expansion of the ostium-meatal complex. Obstruction of the bilateral frontal recesses and mucosal thickening with partial obstruction and re-modeling of the right ostium-meatal complex as well as a soft tissue component that extended from the left alveolar recess into the oral cavity (Figure 3).

Figure 1: Extraoral photography shows increase in volume in the middle third of the left side of the face.

Figure 2: Intraoral lesion in the hard palate.

Figure 3: Computed tomography. The total occupation of the left maxillary sinus was observed.

Following the stablished protocols of our Unit, an incisional biopsy was chosen as the first step of the treatment plan. The conventional microscopic study revealed an undifferentiated neoplasm, constituted by abundant blood vessels, lined by neoplastic endothelial cells, characterized by the presence of a pleomorphic and hyperchromatic nucleus. This cellular component was organized in the form of small clusters as well as individually (Figure 4).

Figure 4: Malignant endothelial cells. Atypical mitotic figures. H&E 400X.

On immunohistochemistry, the markers for mesenchymal tissue (Vimentin), smooth muscle actin (SMA) and CD34 for vascular differentiation were positive in the neoplastic cells. Likewise, the cell proliferation marker Ki-67 was expressed in 60% of them and thus, a diagnosis of angiosarcoma was issued (Figure 5).

Figure 5: CD34(+). Vascular differentiation.

Due to the diagnosis, the tumor size, the surgical inaccessibility and the impossibility to guarantee free margins, the patient was referred to the medical oncology service, where adjuvant radiotherapy was initiated, consisting of 33 sessions with doses of fractioned 200cGy for a total of 6000cGy. Currently, the patient is still receiving the therapy, and is being evaluated periodically. Depending on the evolution, a salvage surgery will be performed.

This study was approved by the Coromoto Hospital, Venezuela IRB and the patient's representatives signed an informed consent agreement.

Discussion and Conclusion

Angiosarcomas represent truly unusual and aggressive malignancies originating from the vascular or lymphatic endothelium. It is an easily infiltrating tumor with a high rate of local recurrence and metastasis [6]. To the authors' knowledge, only 24 cases of this entity in the sinonasal region have been reported in the literature since 1974 [7]. Its appearance has been reported in a wide range of ages ranging from 8 to 81 years, with a frequent peak of onset at 73 years, being more common in males [8]. All these data are in agreement with our case.

The etiology of angiosarcomas is not entirely clear, however, some authors state that its appearance may be related to a history of radiation and lymphedema, immunosuppression and arteriovenous fistulas [9,10]. In the present case, the patient had a significant risk of occupational exposure in the oil industry for a prolonged period of time, in correlation with that described by Tran., *et al*. [11], who claimed that substances such as arsenic, vinyl chloride and torotrast are considered high risk for the appearance of these entities.

On the other hand, some genetic alterations such as bilateral Retinoblastoma, Recklinghausen's Neurofibromatosis, Ollier's disease, Maffuci's disease, Xeroderma Pigmentosa and Klippel-Trenaunay syndrome have been associated with the development of angiosarcomas [12].

Angiosarcomas can occur anywhere on the body, with the skin region being the most common site of appearance (60% of cases). At the level of the head and neck region, although rare, it most often affects the scalp [13,14]. In the sinonasal region, they constitute less than 0.1% of malignant neoplasms [15].

The clinical diagnosis of angiosarcoma might be considered a challenge, due to the nonspecific clinical characteristics of this entity. They may present as single or multiple reddish or purplish nodules, often ulcerating or bleeding. Regarding sinonasal angiosarcomas, the most frequently presented symptoms are epistaxis, obstruction and nasal discharge [16,17].

Differential diagnoses for angiosarcomas include hemangioma, Masson's disease or intravascular papillary endothelial hyperplasia, juvenile nasopharyngeal angiofibroma, hemangiopericytoma, Kaposi's sarcoma and malignant melanoma [18].

On computed tomography imaging, this tumor may present variable dimensions. Several lesions have been reported with dimensions between 2 and 8 cm, with heterogeneous contrast enhancement and signs of bone destruction [7]. In the present case, the imaging characteristics were similar to those previously described.

Microscopically, angiosarcomas are neoplasms that are difficult to differentiate; from well-developed anastomosing vascular channels lined by atypical endothelial cells up to solid sheets of epithelioid or spindle cells may be evidenced. Hemorrhage and necrosis

are common [19]. The vascular phenotype is confirmed by immunohistochemistry, where vascular markers including CD34, CD31, von Willebrand factor (vWF), Fli1 and ERG are expressed. D2-40 might also be present. Due to the varying rates of sensitivity and specificity, a panel of vascular markers is generally recommended in the evaluation of a potential angiosarcoma. CD31 is very sensitive, shows good specificity and is considered by many to be the gold standard for the diagnosis of angiosarcoma [20]. All these findings are also in agreement with the case described.

Due to the limited literature regarding angiosarcomas, there is no standardized treatment. Surgery with safety margins is the treatment of choice, according to the location of the tumor and its extension, followed by adjuvant radiotherapy. Other treatment modalities such as chemotherapy, radiosurgery and the use of interleukins have also been described [21,22]. In our case, due to the large size of the tumor and its location, the multidisciplinary team decided to use radiotherapy as the initial treatment.

The prognosis of angiosarcomas varies according to the degree of differentiation of the tumor and the time of diagnosis. Reported survival rates are 22%. Distant metastasis usually occurs within 24 months in 30% of cases, with the lungs and liver being the sites with the greatest predilection [23].

Due to the unusual nature of this entity, a greater number of reports and reviews on the subject are necessary to establish a management algorithm that allows to improve the prognosis of this pathology. Nevertheless, surgery remains the gold standard for this type of tumor.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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Volume 3 Issue 7 July 2021