PRIMARY EPITHLELIOID ANGIOSARCOMA OF THE MAXILLA MASQUERADING AS A POORLY-DIFFERENTIATED CARCINOMA : CASE REPORT

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Abstract

Epithlelioid angiosarcoma (EA) is a rare high grade malignant neoplasm arising from vascular endothelium. Amongst epithelioid endothelial cell tumors, EA is rare to occur. It is characterized by atypical, multilayered or solid endothelial proliferation and vasoformative architecture is a major finding in EA. This tumor exhibits a great degree of nuclear pleomorphism and mitotic activity with areas of necrosis. A case report of Primary Epithlelioid Angiosarcoma of the Maxilla in a 13 year old male patient is being reported.

Key words: Angiosarcomas, Sarcoma, Primary Epitheliod Angiosarcomas

Introduction

Angiosarcomas are rare malignant tumors of the vascular endothelium, characterized by the formation of irregular vascular channels lined by atypical endothelial cells.[1] They comprise only 2% of soft tissue sarcomas. These tumors arise within bone with an incidence rate of about 0.5%.[2] In the head and neck area, most of these lesions affect the scalp and the facial soft tissues, and only uncommonly, the oral cavity is the site of primary location.[3, 4] Most of the intraoral tumors are located in the mandible and occurrence in the maxilla and the maxillary sinus is rare.[5]

Case Report

A 13-year-old male patient, with a moderate built, yet poorly nourished, reported to the department with a complaint of a swelling associated with pain in the right front region of the upper jaw with inability to close the mouth since 20 days. He gave a history of difficulty in breathing through the right nostril.

Upon detailed extra oral examination, the child had a diffuse mid- facial swelling on the right side. Intraoral examination revealed an irregular but well-defined, sessile growth, The surface was smooth, with areas of hemorrhage and ulceration, presumably resultant to incessant occlusal trauma (fig. 1). It measured 6cms x 7cms in dimension and was confined to the buccal vestibule and alveolar region of the maxilla, extending from the right permanent maxillary lateral incisor to the right permanent maxillary first molar, with extension beyond the mid-palatal raphe. There was shift in the maxillary dental mid-line with marked displacement of the deciduous canine and second molar; The 1st right premolar of the patient was embedded almost completely within the swelling.

The inspection findings were confirmed by palpation. There was a severe tenderness with profuse bleeding on palpating the area; The skin overlying the

maxillary sinus was warm and tender. However, we failed to notice any infraorbital nerve paraesthesia or anesthesia.

A differential diagnosis of sarcoma, aggressive odontogenic tumor and sqamous cell carcinoma were made at the conclusion of examination. Conventional and advanced imaging investigations revealed a diffuse increase in attenuation of maxilla with evidence of osteolytic and destructive lesion. Severe expansion of maxilla and displacement of lateral incisor, canine and premolars were noticed. (Fig. 2, 3) Under local anesthesia, an incision biopsy was performed which concluded a poorly-differentiated carcinoma. Under general anesthesia a wide local resection (subtotal maxillectomy) of the tumor was done. (Fig. 4) The oncologic defect, however, was not reconstructed and was allowed to heal by secondary intention. (Fig. 5) Neck dissection was not contemplated at surgery, but instead a wait-and-watch policy was adopted. The patient's recovery was uneventful. Later, an obturator was fabricated to obturate the defect, facilitate speech and restore masticatory function. The resected specimen was subjected to histopathological examination and a final diagnosis of poorly-differentiated carcinoma was made. (Fig. 6)

Immunohistochemical analysis was performed on the tumor to study its immunoreactivity and various tumor markers were elaborated. The tumor cells show cytoplasmic positivity for vimentin and CD 31 with rare cells showing positivity for CD 34. (Fig. 7,8) At this stage, a definitive diagnosis of epithelioid angiosarcoma was made. In view of the reported resistance of the tumor to radiotherapy, the patient was commenced on a course of post-operative chemotherapy consisting of three cycles each of Vincristine (2mg x3) and Methotrexate (50mg x3). The postoperative period was uneventful. The patient was advised to visit regularly for examination. After 6 months of follow-up, no recurrence was seen.

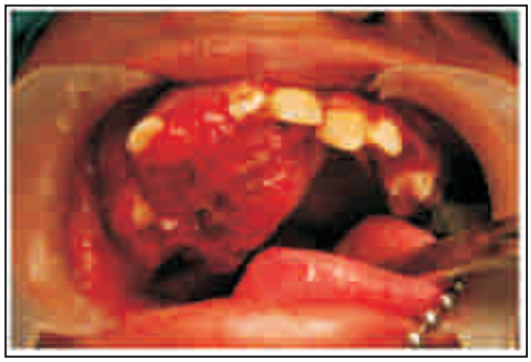

Fig 1 – Intraoral view showing irregular sessile growth, with areas of hemorrhage and ulceration.

Fig 2 – Maxillary occlusal radiograph showing irregular radilucent area with teeth floating in air appearance

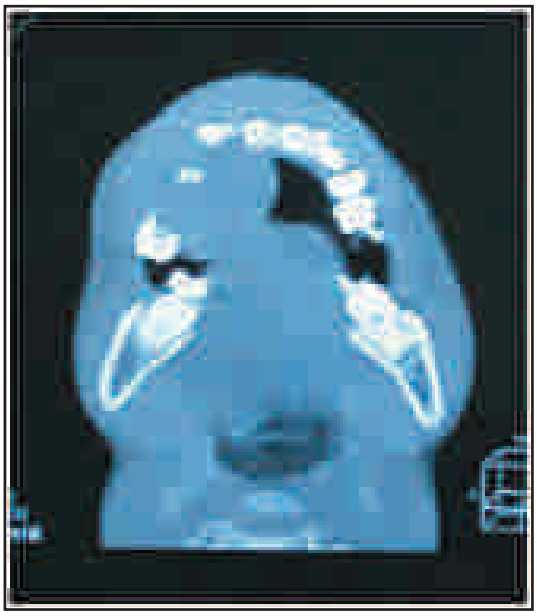

Fig 3 – C T image – right side axial view

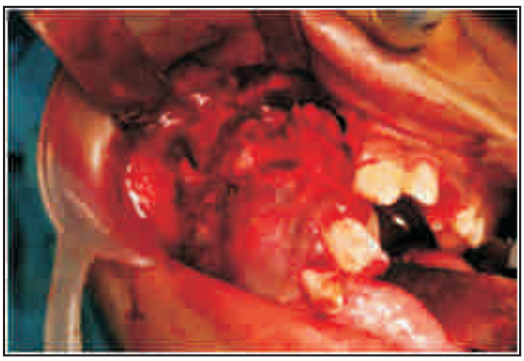

Fig 4 - Intraoperative Photograph

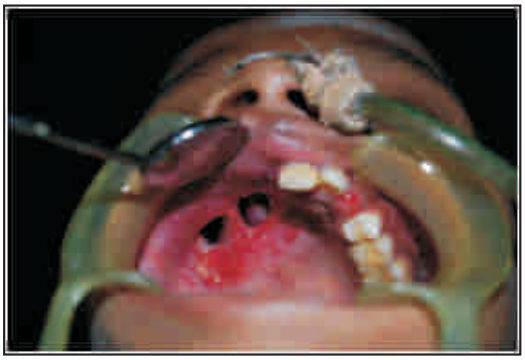

Fig 5 – postoperative photograph

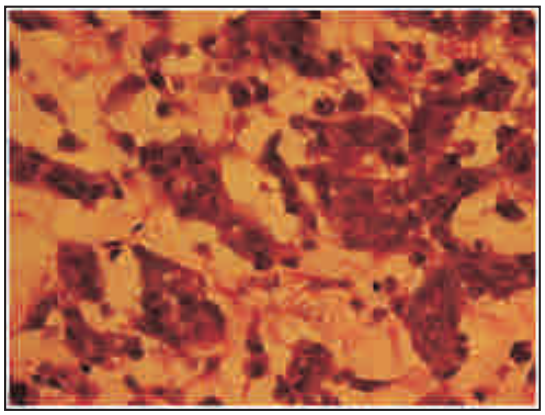

Fig 6 – H & E staining - Epithelioid cells show features like large, pleomorphic vesicular nuclei, many exhibiting nucleoli, and typical and atypical mitosis.

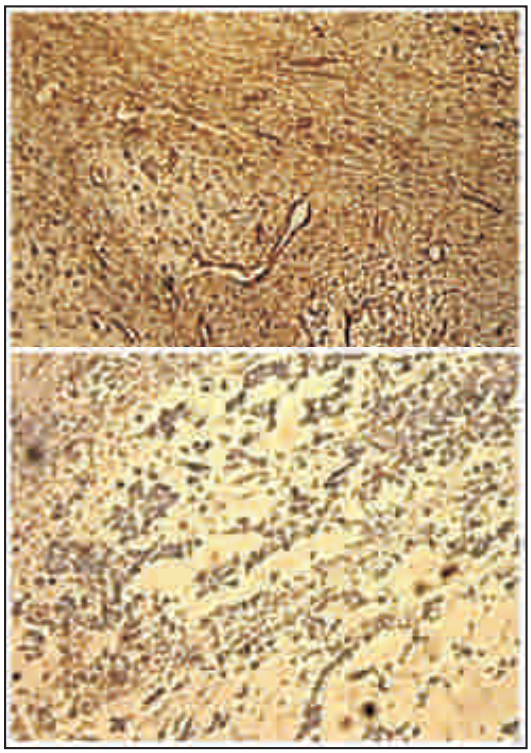

Fig 7 & 8 – IHC - Tumor cell shows cytoplasmic positivity for vimentin and CD 31 with rare cells showing positivity for CD 34

Discussion

Epithelioid angiosarcoma (EA) is a rare malignant tumor of the vascular endothelial cells. The classification of epithelioid endothelial cell tumors includes epithelioid hemangioma,epithelioid hemangioendothelioma and epithelioid angiosarcoma[6,7,8].

Most patients with maxillary angiosarcoma have as short a history of 4 to 8 weeks to being asymptomatic as long as 3 years before diagnosis[4]. The tumor is reported to have no age group, gender or nationality predilection. However, the greatest incidence rate occurs within the fifth decade of life.[5] Tumors in the maxilla or the maxillary sinus produce symptoms related to invasion of surrounding structures, including the eye, orbit and the lateral wall of the nasal cavity. Other reported symptoms include headache, tooth mobility, nasal obstruction, facial asymmetry and diplopia. The present case had

similar symptoms with short duration of swelling.

Many theories have been proposed to explain the development of angiosarcomas. Mc Carthy and Park[6] hypothesized that these tumors arise from the capillaries of granulation tissue in traumatized areas. The same authors have reported 3 patients whose previously benign angiomas underwent malignant transformation after radiotherapy. Vinyl chloride has been implicated as a possible etiologic factor in 30 patients with angiosarcoma of the liver, as reported by Duck[7] in 1975. Williamson and Ramsden, in 1988, have also reported of a 48-year-old man who had a history of handling or being exposed to the chemical for 6 years before an angiosarcoma of the maxillary sinus developed.

Clinically, oral lesions usually appear as painless, sessile-based masses, soft and compressible to moderately firm in consistency and bleed spontaneously, similar to present case. The surface may be ulcerated and an erythematous ring is often present around its periphery.[8,9]

A variety of lesions may be considered in the differential diagnosis, including hemangioma, pyogenic granuloma, papillary endothelial hyperplasia, hemangiopericytoma, angiolymphoid hyperplasia with eosinophilia, Kaposi's sarcoma, malignant melanoma, metastatic disease (renal cell carcinoma) and fibrosarcoma and histiocytic lymphoma in some instances[1,9,10]

The radiographic appearance of angiosarcomas involving the facial bones is generally that of a destructive osteolytic lesion with mild periosteal reaction sometimes mimicking benign lesions such as a cyst or odontogenic tumor. Jaw lesions may show widening of the periodontal ligament and erosion of the alveolar bone producing a "teeth floating in space" appearance.[4]

Histopathological diagnosis, particularly with poorly differentiated angiosarcomas and the epithelioid phenotypic variant of the tumor may be confused with spindle cell sarcomas, carcinomas or a melanoma. Previously, tumor markers like factor VIII-related antigen and ulex europeus agglutinin lectin were used to assist in immunohistochemical diagnosis. However recently, the vascular and lymphatic endothelial expression of another antigen, CD31, has been proven to be the most sensitive endothelial marker[11,12]. CD 31 yields strong labeling of the lesional cells with low background immunoreactivity. Difficulties with background staining have plagued the use of lectin while factor VIII-related antigen although specific, has low sensitivity.

As far as treatment is concerned, most authors believe that surgery, in combination with radiotherapy and chemotherapy, offers the best chance of survival[8]. Zachariades and Economopoulou, however, have advocated treatment consisting of wide surgical removal of the tumor regardless of the combination of irradiation[8].

EA, as a whole, have a strong tendency to recur locally and to metastasize indicating their highly aggressive nature which explains the low survival rates. McCarthy and Park reported a 3-year survival rate of 17% and a 5-year survival rate of 9% with an average survival time of 2½ years. The survival rate for oral and jaw angiosarcomas, however, is better than that for angiosarcomas in general and, in particular, for those involving the skin. Many investigations in recent years have indicated that one-half of the patients with angiosarcomas die within 15 months from the time of initial diagnosis, with approximately 12% surviving 5 years or longer.[11,12]

Conclusion

The frequency of Angiosarcomas in the maxillofacial region is exceedingly sporadic. Since Angiosarcomas offer a grave to poor prognosis in the long run, a more in depth understanding of the behavior and

genetics of these tumors is paramount to their better management in the future.

Reference

1. *Bankaki M, Myers EN, Barnes L, DuBois P. Angiosarcoma of the maxillary sinus: literature review and case report. Head Neck Surg 1979; 1:274-80.*
2. *Enzinger FM,Weiss SW; Soft tissue tumors. St LouisMosby, 1983 p 430.*
3. *Zachariades N, Economopoulou P. Maxillary angiosarcoma. Int 1 J Oral Maxillofac Surg 1986; 15:357-60.*
4. *Aust MR, Olsen KD, Lewis JE, Nascimento AG, Meland NB Foote RL, et al. Angiosarcomas of the head and neck: clinical and pathologic characteristics. Ann Otol Rhino Laryngol 1997 106:943-51.*
5. *Angiosarcoma of maxilla and maxillary sinus; JOMS; 1989; 47; 747-753.*
6. *McCarthy WD, Pack GT. Malignant blood vessel tumors. A report of 56 cases of angiosarcoma and Kaposi's sarcoma. Surg Gynecol Obstetric 1950; 91:465-82.*
7. *Mark RJ, Poen JC, Tran LM, Fu YS, Juillard GF. Angiosarcoma. A report of 67 patients and a review of the literature. Cancer 1996; 77:2400-6.*
8. *Zachariades N, Papadakou A, Koundouris J, Constandinidis J, Angelopoulos AP. Primary hemangioendotheliosarcoma of the mandible: review of the literature and report of a case. J Oral Surg 1980; 38:288-96.*
9. *Haustein UF. Angiosarcoma of the face and scalp. Int J Dermato 1991; 30:851-6.*
10. *Loudon J A, Billy ML, De Young BR, Allen CM. Angiosarcoma coma of the mandible: a case report and review of the literature Oral Surg Oral Med Oral Pathol Oral Radiol Endoc 2000;89:471-6.*
11. *Epitheloid angiosarcoma of maxillary sinus and maxilla; A case report and review of literature; Oral Surg,Oral Med Oral Pathol Oral Radiol Endo2002;94:333-337.*
12. *Kempson RL, Fletcher CD, Evans HL, Hendrickson MR, Sibley RK. Atlas of tumor pathology. Tumors of the soft tissues. Washington, DC: Armed Forses Institute of Pathology; 2001. p. 353.*

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