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Case Report

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A case of angiosarcoma of the nasal cavity

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ABSTRACT

Angiosarcoma of the nasal cavity is extremely uncommon accounting for less than 0.1% of all sinonasal malignancies. Here we report a case of angiosarcoma of nasal cavity. A 45 year old male patient with c/o right sided nasal bleeding since 20days, on examination showed mass in nasal cavity. Patient underwent septoplasty with right inferior turbinectomy with submucosal diathermy. The histological examination showed features of angiosarcoma epitheloid variation of nasal cavity, diagnosis was confirmed by immunohistological stain with factor CD 31 and cytokeratin.

Keywords: Angiosarcoma, Nasal cavity, Septoplasty, Turbinectomy, Submucosal diathermy, CD 31

INTRODUCTION

Angiosarcomas are malignant neoplasm of rapid growth that develop from endothelial cells. They represent 2% of all sarcomas and only 1-4% are located in the aerodigestive tract. Angiosarcomas are extremely rare in the head and neck.² Angiosarcoma is an uncommon malignant neoplasm characterized proliferating, extensively infiltrating anaplastic cells derived from blood vessels and lining irregular bloodfilled spaces. Angiosarcomas may occur in any region of the body but are more frequent in skin and soft tissue. Angiosarcomas also can originate in the liver, spleen, bone, or heart, breast, uncommon in the sinonasal region (<0.1% of all sinonasal malignancies). Sites of involvement in head and neck region include the scalp and forehead, cheek, nose and ethmoid sinuses, neck, and mandible.³ They appear during middle age and prognosis depends on location, size and degree of tissue invasion.¹ Since 1977 only 17 cases have been reported in literature.4

CASE REPORT

A 45 year old male patient with c/o Right sided nasal bleeding since 20 days. Diagnostic nasal endoscopy shows right side a reddish friable mass 1cm X 1cm extending from medial surface of inferior turbinate to septum, scope could not be passed beyond the mass to visualize nasopharynx. Middle meatus found to be clear. No neck lymph nodes enlargement. CT PNS scan showed minimal mucosal thickening in B/L maxillary sinuses and non-enhancing soft tissue density extending from right inferior turbinate to nasal septum (Figure 1 and 2).

Patient underwent septoplasty with right inferior turbinectomy with submucosal diathermy and specimen sent for histopathology.

Microscopic review of the specimen revealed a vascular neoplasm consisting of vascular spaces of various calibre with lining atypical endothelial cells with rounded moderately pleomorphic nuclei having prominent nucleoli with intacytoplasmic vacuoles (neolumen formation) containing RBCs and diagnosed as Angiosarcoma epitheloid variation of nasal cavity (Figure 3 A and B). Diagnosis confirmed by immune-histochemical markers – CD 31 strongly positive and cytokeratin was negative. PET scan was done. PET scan showed no signs of metastasis. Patient refused radiotherapy (Figure 4).



Figure 1: CT scan axial cuts showing tumour in right nasal cavity extending between septum and inferior turbinate.



Figure 2: CT scan coronal cuts showing tumour in right nasal cavity.

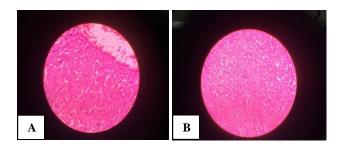


Figure 3 (A and B): Microscopic picture of angiosarcoma of the nasal cavity.



Figure 4: DNE showing no recurrence 1 year postsurgery.

DISCUSSION

Angiosarcomas are extremely rare in the head and neck.² Etiology is unknown and has been associated to certain risk factors such as chronic lymphedema, radiotherapy, vinyl chloride exposure, trauma and telangiectatic skin lesions. Sites of involvement included the scalp and forehead, cheek, nose and ethmoid sinuses, neck, and mandible.³ The principle differential diagnosis includes granulation tissue, lobular capillary hemangioma (pyogenic granuloma), and Kaposi's sarcoma. Diagnosis of angiosarcoma is established by pathological examination of the biopsy using H&E. However, immunohistochemical confirmed by staining.1 Agiosarcomas of the upper aerodigestive tract have a better prognosis and long term survival free of disease than their soft tissue and skin counterparts. Recurrence is strongly correlated with positive resection margins. There is no standard treatment for nasal cavity and paranasal sinus angiosarcoma. Complete surgery and radiotherapy are considered the optimal treatment schemes.⁵ Kazuto Fukushima et al. reported favorable results with the use of recombinant interleukin 2, combined with surgery.⁶ Angiosarcoma of the skin or soft tissue of the head and neck is associated with a 50% mortality rate within the first 25 months and a 12% survival rate at 5 years, compared to nasal cavity or paranasal sinus angiosarcoma, which have a 22% survival rate at 5 years according to grade of differentiation and early diagnosis.¹

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