International Journal of Otorhinolaryngology and Head and Neck Surgery
Singh J et al. Int J Otorhinolaryngol Head Neck Surg. 2017 Jul;3(3):735-736
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

Case Report

DOI: http://dx.doi.org/10.18203/issn.2454-5929.ijohns20173058

Angiosarcoma of the nasal cavity and paranasal sinuses: a rare case report

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Received: 28 September 2016
Accepted: 02 November 2016

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ABSTRACT

Angiosarcomas are the malignant neoplasias of vascular phenotype whose constituent tumor cells have endothelial features. They account for less than 1% of all sinonasal tract malignancies. A case of 65 year old male with a 6 months history of spontaneous epistaxis and a nasal mass lesion presented to the OPD. On physical examination, a polypoidal purplish tumor of 6 × 5 cm was identified. A NCCT scan of nose and paranasal sinuses showed a nonenhanced tumor in the left nostril in maxillary, ethmoid, sphenoid and frontal sinus. An excisional biopsy revealed a poorly differentiated angiosarcoma. Surgical removal of the lesion was advised which was refused by the patient. Patient was put on the radiation therapy. Angiosarcoma of the nose and paranasal sinuses is an extremely rare tumor. Recurrences are common due to incomplete excision or mutiflocality.

Keywords: Angiosarcomas, Nasal cavity, Paranasal sinuses

INTRODUCTION

Angiosarcomas are high-grade, malignant vascular tumors that make up only about 1% of all sarcomas.[1,2] They develop from endothelial cells. Primary sinonasal tract angiosarcomas are exceedingly uncommon and only a few cases have been reported in the english literature.[3]

Since 1977 only 17 cases have been reported in literature. They present as a mass lesion with or without epistaxis and airway obstruction.[4] The principle differential diagnosis includes granulation tissue, lobular capillary hemangioma (pyogenic granuloma), and Kaposi's sarcoma. Complete surgical excision is the treatment of choice. Radiotherapy and chemotherapy may be beneficial too.

CASE REPORT

A 65-year-old male patient presented with 6 months history of nasal mass in the left nasal cavity, spontaneous epistaxis with occasional headache. During physical examination, there is polypoid mass seen occupying whole of the left nasal cavity. There is no active anterior bleeding. A NCCT scan of paranasal sinus shows a nonenhanced tumor in the left nostril in maxillary, ethmoid, sphenoid and frontal sinus measuring 5 cm, 5.5 cm, and 6 cm, Figure 1.

Histopathological examination of specimen demonstrated poorly differentiated angiosarcoma. The microscopic view showed anastomosing vascular channels lined by remarkably atypical endothelial cells protruding into the lumen, neolumen formation, frequent atypical mitotic figures, necrosis, and hemorrhage.

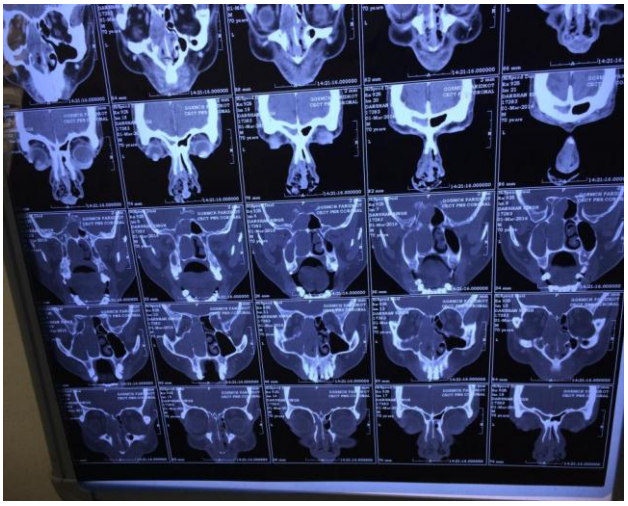

Figure 1: NCCT Nose and PNS.

Excisional biopsy was taken from the mass which showed malignant mesenchymal tumor.

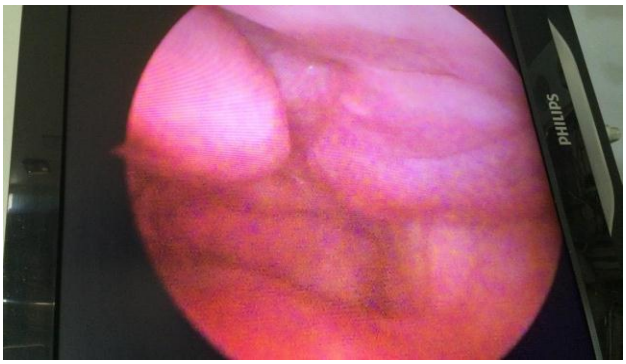

Figure 2: Endoscopic image of the patient (post excision)

On immunohistochemistry, the tumour cells showed diffuse strong cytoplasmic membrane positivity with CD31 and are positive for CD34, non-reactive to keratin and S 100. Patient was advised to undergo total maxillectomy, but refused for the surgery and was referred to the radiotherapy department. Patient remains on follow up 3monthly.

DISCUSSION

Excisional biopsy was taken from the mass which showed malignant mesenchymal tumor.

Histopathological examination of specimen demonstrated poorly differentiated angiosarcoma. The microscopic view showed anastomosing vascular channels lined by remarkably atypical endothelial cells protruding into the lumen, neolumen formation, frequent atypical mitotic figures, necrosis, and hemorrhage.

On immunohistochemistry, the tumour cells showed diffuse strong cytoplasmic membrane positivity with CD31 and are positive for CD34, non-reactive to keratin and S 100.

Angiosarcomas are malignant neoplasias of rapid growth that originate from endothelial cells.[1,2] They represent 1% of all sarcomas, and primary sinonasal tract angiosarcoma is 0.1%.[3] They appear during middle age, more common in males than females and prognosis depends on location, size and degree of tissue invasion. Patients present with chief complaints of recurrent epistaxis, nasal blockage. Etiology is unknown and known to have the risk factors like chronic lymphedema, radiotherapy, vinyl chloride exposure, trauma and telangiectatic skin lesions.[5,6] Diagnosis is made by histopathological examination of the biopsy from the mass and confirmation by immunohistochemical stains which shows positivity to CD 34, CD 31,Ulex europaeus agglutinin I and factor VIII antigen. Treatment of choice is the radical surgical excision of the mass with free margins followed by radiotherapy. Recurrences are common due to incomplete excision. Prognosis of angiosarcoma of sinonasal tract is better than angiosarcoma of any other site.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Singh J, Davessar JL, Prinja S, Arora N. Angiosarcoma of the nasal cavity and paranasal sinuses: a rare case report. Int J Otorhinolaryngol Head Neck Surg 2017;3:735-6.