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Epithelioid hemangioendothelioma of the nasal septum

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Abstract. *Epithelioid hemangioendothelioma of the nasal septum. Background*: Epithelioid hemangioendothelioma is a tumour of vascular origin and unknown aetiology, which occurs in all age groups. The most common locations are the liver, bone, lungs, and brain, with less common occurrence in head and neck regions, and no prior reports of origination from nasal septum.
Case report: Here we report a case of epithelioid hemangioendothelioma of the nasal septum. A 62-year-old male patient attended our clinic with epistaxis and congestion in his left nasal cavity. Endoscopic examination revealed a mass within the left nasal passage. Clinical and histopathological examinations showed that the mass was a septum-originated epithelioid hemangioendothelioma. The mass was removed endoscopically using a transnasal approach.
Conclusions: No recurrence was observed over 36 months of follow-up. Here we discussed this uncommon case along with a literature review.

Introduction

Epithelioid hemangioendothelioma (EH) is a low-grade tumour that was first described as a vascular neoplasm of endothelial origin in 1982 by Weiss and Enzinger.[1,2] This vascular neoplasm has been reported to occur in various organs and locations, including soft tissue, bone, skin, lymph nodes, brain, liver, spleen, colon, peritoneum, lung, pleura, heart, the vascular system, and rarely in the head and neck.[1,3] EH treatment usually involves wide surgical excision of primary tumours if there are no signs of metastasis, with clinical follow-up due to the risk of recurrence.[3,4] Here we report the first case of EH arising from nasal septum, and we discuss this case with a review of the literature.

Case report

A 62-year-old male patient arrived at our clinic presenting congestion in his left nasal cavity and recurrent epistaxis for one month. His medical history was not relevant, except his smoking habit of 20 packets/year. Physical examination found a painless, ulcerated, bleeding tumour with a diameter of 2 cm in the left side of the nasal cavity. Endoscopic examination revealed that the mass appeared haemorrhagic and ulcero-vegetant (Figure 1A). Paranasal computerized tomography (CT) showed a suspicious mass lesion compatible with soft tissue density, located in the left nasal cavity, situated between the left inferior and middle turbinate, and thought to stem from the septum (Figure 1B). Immunohistochemical examination via punch biopsy revealed that $CD31^+$, $CD34^+$, SMA^+, and Ki-67expression level was around 5% and the patient was diagnosed with a low-grade EH (Figure 1C,D).

Results

Based on these findings, we planned endoscopic surgery. During surgery, the haemorrhagic mass of the septum was widely excised, including septum mucosa, perichondrium, and cartilage. Microscopic examination of the specimen confirmed the diagnosis of EH. We observed no signs of recurrence during 36 months of patient follow-up.

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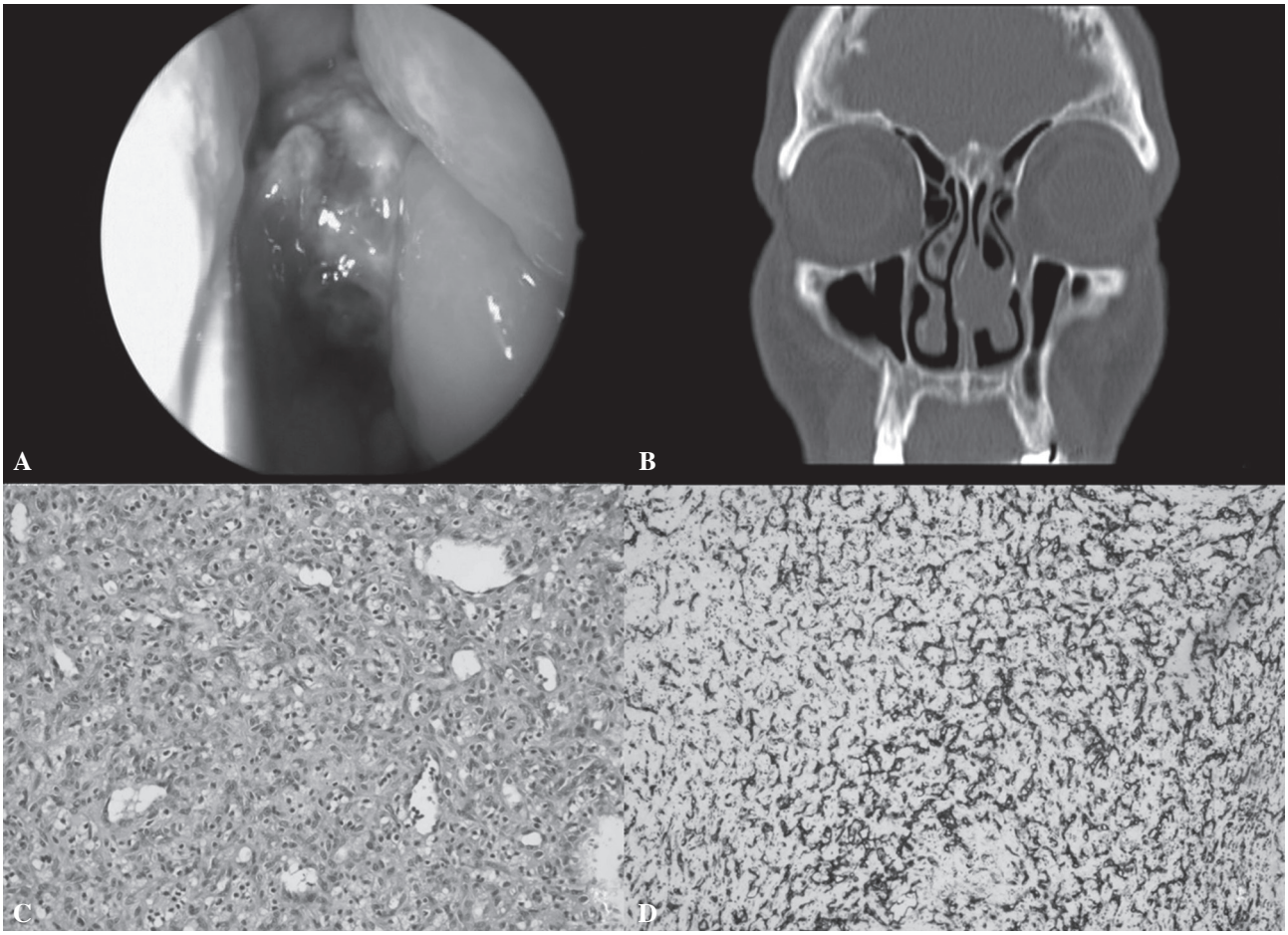

Figure 1
(A) Left nasal endoscopy showing a haemorrhagic and ulcero-vegetant mass. (B) Computerized tomography (CT) image showing a suspicious mass lesion with soft tissue density in the left nasal cavity. (C) Hematoxylin-eosin staining reveals atypical endothelial cell proliferation on myxoid ground (20× magnification). (D) Tumour cells show strong immunostaining with CD34 antibody.

Discussion

EH is a low-grade vascular neoplasm of endothelial origin, for which the aetiology is not yet completely understood.[1] EH is more frequently observed on soft tissue, bone, skin, lymph nodes, brain, liver, spleen, colon, peritoneum, lung, heart, and the vascular system – but is also rarely detected in regions of the head and neck.[1, 2] Here we report a case of EH of the nasal septum, which has not been described in any previous report in the literature.

EH can occur in patients of any age, and its frequency is the same for both sexes. Predisposing factors have not yet been identified; however, EH is reportedly associated with trauma, therapeutic radiation, and hormonal factors.[2] Clinically, it is challenging to distinguish nasal EH from benign lesions, such as pyogenic granuloma, haemangioma, angiomyoma, arteriovenous malformation, and hamartoma. Definitive diagnosis relies on analysis of histopathological and immunohistochemical features. For microscopic definitive diagnosis, one must also consider metastatic carcinoma, epithelioid angiosarcoma, and epithelioid sarcoma.[2]

Tumour cells can form painful masses on superficial or deep tissue. Upon inspection, a white and red appearance indicates the mass vascularity.[1] Immunohistochemical analyses may yield reliable clues regarding differentiation. Tumour cells can display mesenchymal or endothelial characteristics. Mesenchymal-like cells show positive staining for vimentin, while endothelial-like tumour cells will stain positive for factor VIII-related antigen, CD34, CD31, and UEA-1.[5,6] Negative staining results for cytokeratin and EMA can rule out pathologies of epithelial cell origin. Similarly, negative staining

for S-100 protein rules out pathologies of Schwann cells, cartilage, and adipose tissue.[7]

In our present case, immunohistochemical examination via punch biopsy revealed that $CD31^+$, $CD34^+$, SMA^+ and Ki-67 expression level was around 5% . Accordingly, the patient was diagnosed with a low-grade EH.

While the prognosis of EH on regions of the head and neck is not completely known, poor prognosis is generally associated with advanced cellular atypia, high mitotic activity, and presence of necrosis.[2] Our reported case showed mild cellular atypia and low mitotic activity, with no necrosis. Thus, the lesion was regarded as borderline, with biological features between those of an angiosarcoma and a haemangioma. Wide local excision with endoscopic endonasal surgery is an approved treatment for EH, which is most often curative.[8,9] In some cases, radiotherapy and chemotherapy have been used as treatment methods.[10] For histologically malign EH, radical local excision is recommended, as is suggested for other sarcomas. One prior study included a 4-year follow-up of an EH series, and reported local recurrence in 13% of the patients, metastasis in 31% of the patients, and 13% mortality.[8]

In our present case, we did not apply chemotherapy or add-on therapy because the surgical margins were free of tumour following complete excision of the mass together with septum mucosa, perichondrium, and cartilage. The patient showed no signs of recurrence or metastasis during 36 months of follow-up.

Conclusion

In conclusion, EH is an infrequently observed tumour of vascular origin. It is rarely detected in regions of the head and neck, with very few reported cases in the nasal cavity. Definitive diagnosis should be supported with clinical examination, imaging, and histopathological and immunohistochemical investigations. We hope that this first report of EH emerging from the nasal septum will increase awareness of this uncommon tumour as a possible diagnosis in cases of nasal septum masses.

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