



CASE STUDY

Epithelioid haemangioendothelioma of the nasal cavity in a woman



Hemangioendoteloma epitelioido de la cavidad nasal en una mujer

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Case report

A 32-year-old woman presented with a 7-month history of intermittent left-sided epistaxis and watery rhinorrhea without seasonal predominance. Her medical history included a septorhinoplasty 3 years before. No bleeding source was identified on repeated nasal endoscopies. There were no signs to clarify the source of bleeding on CT, although a coronal cut between the septum and the middle third of the middle turbinate showed an image that was similar to a drop of liquid (Fig. 1). A more thorough endoscopic exploration under general anesthesia and, if needed, sphenopalatine artery bipolar coagulation was considered. A small tumor was found in the medial side of the vertical portion of the middle turbinate body, facing the septum. An endoscopic resection of the vertical aspect of middle and superior turbinates that included a 1-cm bluish-purple tumor in the septal aspect of the left middle turbinate was performed. The pathology report was "EH 0.9 cm in maximum diameter with negative surgical

margins. Immunohistochemically, tumor cells are immunoreactive for vimentin, CD31, CD34, α -1-antichymotrypsin, and FVIII" (Figs. 2 and 3). At 18 months follow-up, the patient remained free of symptoms and nasal endoscopy showed no abnormalities.

Discussion

EH was first described by Weiss and Enzinger in 1982 as a tumor that affects soft tissue.¹ It is an angiocentric vascular tumor with endothelial component. Both by clinical behavior as by histological characteristics, EH is considered a tumor of intermediate malignancy because, although it has potential to recur or metastasize, it does so with a frequency and speed much lower than those of conventional angiosarcoma. It appears at any age and is often closely associated to a blood vessel, usually a medium-sized or large vein.² It can occur in several locations: soft tissue, bone, liver, lung, skin, oral cavity, gastrointestinal tract, peritoneum, lymph nodes, meninges, mediastinum, pleura, heart, spleen, thyroid, or parotid gland.³ Only six cases of this tumor in the nasal cavity have been found up to 2016 in the review of literature in English carried out (Table 1).^{3–8} The tumor consists of endothelial cells with epithelioid and

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Figure 1 Coronal section of unenhanced CT: soft tissue window. Mucosal thickening and slight hypodensity of the septal mucosa of the left middle turbinate (arrow).

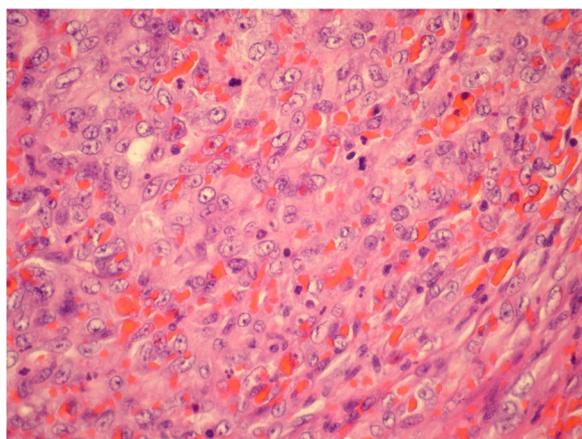


Figure 2 Histologic image from resected tumor. Tumor is composed of solid nests of large cells with abundant cytoplasm (epithelioid), and large nuclei with clear nucleolus. Two cells are in mitosis. There are abundant small vascular channels formed by tumor cells and intracytoplasmic lumina containing erythrocytes. Hematoxylin–eosin, original magnification $\times 400$.

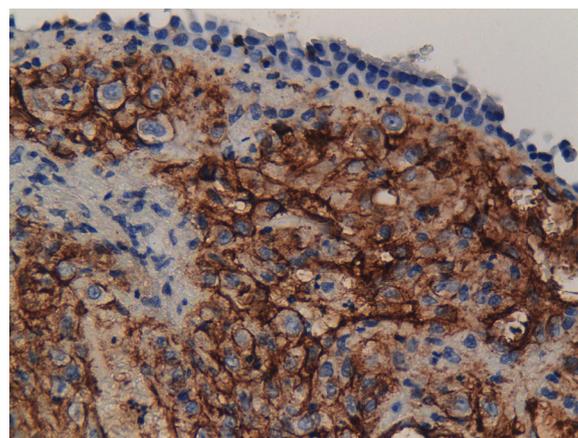


Figure 3 Immunohistochemistry from resected tumor. CD31. Endothelium specific antibody. Positive membrane staining in tumor cells. Respiratory epithelium is visible on the surface, which is negative. Original magnification $\times 40$.

Table 1 Features of present case and previously reported cases of epithelioid hemangioendothelioma of the nasal cavity.

<i>n</i>	Sex	Age	Site	Clinic	Treatment	Follow-up	Year	Author
1	M	23	Middle meatus	Intermittent epistaxis	Surgery	12 m NED	2003	Di Girolamo ³
2	M	25	Middle turbinate	Intermittent epistaxis	Surgery	36 m NED	2005	Tseng ⁴
3	M	4	Nasal cavity	NA	Surgery	2 recurrences at 3 and 5 y. Now 12 y NED	2008	Naqvi ⁵
4	M	40	Septum	Intermittent epistaxis	Surgery	9 m NED	2010	Patnayak ⁶
5	M	54	Nasal cavity	Nasal obstruction	Surgery	24 m NED	2012	Hao ⁷
6	F	27	Nasal cavity	Intermittent epistaxis and facial pain	Surgery	26 m NED	2016	Ogita ⁸
7	F	32	Middle turbinate	Intermittent epistaxis	Surgery	18 m NED	2016	Current

Abbreviations: M, male; F, female; m, months; y, years; NA, not available; NED, no evidence of disease.

histiocytoid appearance. It usually appears as a single mass in soft tissue at any anatomic site, but when it does so in the lung, liver, and bone tends to be multifocal. Tumor cells have eosinophilic cytoplasm and rounded nucleus, within a myxoid stroma. Characteristically, the cytoplasm of these cells is vacuolated, which helps establish the diagnosis. It is also typical that these vacuoles harbor erythrocytes and converge to form intracytoplasmic lumina, which could represent aborted vascular channels. Nuclear pleomorphism is minimal and no mitoses are usually observed. Histologically EH may be mistaken for an angiosarcoma, but the latter usually has more cellular atypia, mitoses, and necrosis. The neoplastic cells have immunoreactivity for endothelial markers such as factor VIII-related antigen, *Ulex europaeus* agglutinin 1 lectin, CD31, CD34, vimentin, and FLI1. Wide local excision of tumor with surgical margin is the appropriate treatment, though it requires a close clinical follow-up because of its potential to recur and metastasize.

There are few cases of EH in the head and neck, and the localization of this tumor in the nasal cavity is exceptional. Only six cases have been reported, one of them in a woman. We present the second epithelioid hemangioendothelioma case in the nasal cavity of a woman.

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Conflict of interest

The authors declare that they have no conflict of interest.

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