Case Report

Kaposiform Hemangioendothelioma of Paranasal Sinus

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Kaposiform hemangioendothelioma (KHE) of the paranasal sins (PNS) is a rare cause of recurrent epistaxis. To date, only two cases of PNS KHE have been reported in the literature, both occurring in the pediatric population. The case presented here appears to be the first case of PNS KHE occurring in an adult.

A 46-year-old white female presented with progressively worsening unilateral recurrent epistaxis. Diagnostic histopathology confirmed it to be KHE.

After a detailed workup, the tumor was completely excised en bloc (medial maxillectomy; anterior and posterior ethmoidectomy) via a lateral-rhinotomy approach. Complete excision of the tumor with clear margins offers the best results.

Key Words: Kaposiform hemangioendothelioma; endothelioma; hemangioma; epistaxis; paranasal sinus; nasal cavity.

INTRODUCTION

Kaposiform hemangioendothelioma (KHE) is an uncommon vascular neoplasm of endothelial origin. It is derived from endothelial spindle cells and has features ranging from those of a benign well-differentiated hemangioma to those of an anaplastic angiosarcoma. Therefore, KHE is locally aggressive, does not regress spontaneously, and has a tendency to metastasize locally as well as to regional lymph nodes. According to the latest World Health Organization classification of soft tissue tumors, KHE is regarded as a tumor of intermediate malignant potential.

KHE was first described in 1971 as a “hemangioma with Kaposi’s sarcoma-like features”; however, other names such as Kaposi-like infantile “hemangioendothelioma” or “hemangioendothelioma” have been used in the past to describe this tumor. It was Zuckerberg et al. who first coined the term “Kaposiform hemangioendothelioma” in 1993.

The tumor typically presents in infancy and early childhood, but can occasionally be seen in adolescents and adults as well. The common sites of occurrence are superficial and deep soft tissue of the extremities, trunk, and retroperitoneum. KHE of the paranasal sinuses (PNS) is extremely rare. To date, only two cases of PNS KHE have been reported in the literature, both of them occurring in the pediatric age group. We present a case of KHE arising from the PNS of a middle-aged female. To the best of our knowledge, this is the first case of PNS KHE occurring in an adult.

CASE REPORT

A 46-year-old white female presented to ENT clinic with a 3-month history of progressively worsening, recurrent left-sided epistaxis and hyposmia. There was no history of allergic rhinitis chronic nasal problems, headache, facial discomfort, visual disturbances (diplopia, ophthalmoplegia), epiphora, fever, facial or nasal trauma, or any nasal surgery.

Flexible nasendoscopy revealed a large vascular mass in the middle and upper portion of the left nasal cavity above the middle turbinate, without any polyps or discharge. The rest of the ENT examination was unremarkable.

Diagnostic histopathology confirmed a vascular neoplasm of uncertain behavior. The case was discussed in

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a multidisciplinary head–neck tumor board, and complete excision of the mass was recommended pending a carotid angiogram.

Carotid angiogram revealed an intensely vascular localized tumor in the ethmoidal region and left nasal cavity, with dual blood supply from both the external and internal carotid arteries supplying two- thirds and one-third of the lesion via ethmoidal branches of the maxillary and ethmoidal branches of the ophthalmic arteries, respectively. Complete blood counts and clotting profiles were in the normal range.

The patient underwent medial maxillectomy, anterior and posterior ethmoidectomy, and en bloc excision of the tumor with clear margins via a lateral-rhinotomy approach. No preoperative embolization was performed. The postoperative recovery was uneventful. Histological findings confirmed a vascular tumor with lobular, peritheliomatous, and trabecular growth pattern, which was infiltrating the surrounding respiratory mucosa and bone (Fig. 2). The tumor was composed of slit-like vascular spaces, spindle cells, and collagenous stroma in varying proportions. The spindle cells had moderately pleomorphic vesicular nuclei with mitotic activity. There was a strong expression of CD31, CD34, and actin on immunohistochemistry (Fig. 3), but no staining for HHV8, desmin, S100, or EMA. These features were in keeping with KHE.

The patient remains disease-free up till a 1 year follow-up.

DISCUSSION

Owing to the rarity of PNS KHE, comprehensive details about clinical presentation, progression, and outcomes are lacking. Out of three cases reported thus far (including the case presented here), two patients were in the pediatric age group (4 months; 8 years) and only the case presented here is an adult.7,8 The tumor appears to have a predilection for the female gender because all the three patients were females. The tumors were located in the ethmoid sinus in two patients, whereas in one patient the tumor was centered in the maxillary sinus.

Recurrent epistaxis was the most common presenting symptom and was seen in all three patients. Hypo- smia and nasal obstruction were the presenting complaints in two cases, whereas periorbital, ecchymosis facial, palatal, and gingival swelling was seen in one case.

Unlike the KHE of other sites, PNS KHE did not show any signs of Kasabach-Merritt phenomenon (KMP),3 which is characterized by features of profound thrombocytopenia and consumption coagulopathy, resulting in life-threatening hemorrhage. The laboratory hallmark consists of platelet counts less than $50 \times 10^9/mL$.
The PNS KHE is a rare cause of recurrent epistaxis affecting both pediatric and adult populations. The treatment of choice is surgical excision with clear margins.

**BIBLIOGRAPHY**


**CONCLUSION**

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