CASE REPORT

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KAPOSIFORM HEMANGIOENDOTHELIOMA ARISING IN THE ETHMOID SINUS OF AN 8-YEAR-OLD GIRL WITH SEVERE EPISTAXIS

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Abstract: Background. Epistaxis is very common during childhood. It occurs primarily in boys and is usually self-limiting. Trauma and nose picking are among the most common causes. In general, epistaxis can be easily treated with anterior nasal packing or electrocoagulation.

Methods. We report a case of an 8-year-old girl with severe unilateral epistaxis.

Results. The bleeding originated from a kaposiform hemangioendothelioma arising in the left nasal cavity and ethmoid sinus. The feeding vessels originating from the maxillary artery were first embolized. The tumor was then surgically removed through a combined external ethmoidectomy and endonasal approach. The postoperative course was uneventful. MRI at 6 months after surgery showed no tumor recurrence.

Conclusions. We report a previously undescribed cause of epistaxis in children, namely, a kaposiform hemangioendothelioma. To our knowledge, this is the first such case in the English-language literature. The differential diagnosis of severe unilateral nasal bleeding among the pediatric population should include the possibility of a kaposiform hemangioendothelioma.

Keywords: epistaxis; kaposiform; hemangioendothelioma; embolization; endoscopic endonasal surgery; external ethmoidectomy

Dry nasal mucosa, rhinitis, foreign bodies, trauma, and nose picking cause the vast majority of cases of epistaxis in children. The source of bleeding is usually anterior, and can easily be visualized and treated.

Severe and recurrent bilateral hemorrhage in children should first prompt blood tests to rule out an underlying coagulopathy. Severe unilateral epistaxis, in the absence of bleeding disorders, also raises the possibility of sinonasal neoplasia and must be further investigated. However, most of these tumors are benign. Hemangioma, which is the most common tumor in infancy, is a major cause of epistaxis attributable to neoplasia. Another important cause of bleeding due to vascular tumors is juvenile angiofibroma, occurring almost exclusively in teenage males. Other well-vascularized solid tumors (ie, rhabdomyosarcoma or olfactory neu-
roblastoma) must be included in the differential diagnosis. Congenital midline masses usually do not cause epistaxis.1

We report a rare case of an 8-year-old girl with severe unilateral epistaxis due to a kaposiform hemangioendothelioma.

**CASE REPORT**

A previously healthy 8-year-old girl experienced a severe and sudden left-sided epistaxis as she was lying in bed. An external ear, nose, and throat (ENT) specialist detected a heavily bleeding mass at the level of the middle turbinate and stopped the bleeding by inserting a nasal packing on the left side. The child was then emergently referred to Children’s Hospital for further investigation and management.

On admission, the patient was in stable condition. No nasal bleeding was present at the time, and blood parameters showed a hemoglobin level of 105 g/L without any signs of coagulopathy.

Contrast-enhanced CT scan of the nose and paranasal sinuses revealed a neoplasm in the left nasal cavity and ethmoid sinus (Figure 1A and B). To obtain better resolution of the mass and information regarding its vascularization and to rule out intracranial extension, both MRI and MR-angiography were performed (Figure 2A). These investigations confirmed a more expansive (rather than infiltrating) mass whose origin was suspected to be at the olfactory rim. The tumor did not extend intracranially but protruded into the orbit with lateral displacement of the medial rectus muscle. Radiologically, a vascular malformation or a benign hemangioma seemed rather unlikely; hence, a rhabdomyosarcoma or olfactory neuroblastoma appeared to be the most probable diagnosis.

Ophthalmologic examination showed no impairment of vision or eye motility. Four days after admission to our clinic, the nasal packing was removed with the patient under general anesthesia. Nasal endoscopy revealed a white mass with smooth borders between the middle turbinate and the nasal septum; the mass was covered with a mesh of blood vessels. A biopsy was taken and resulted in massive hemorrhage. Cauterization failed to stop the bleeding, which was finally controlled by inserting nasal packing. The patient’s hemoglobin level dropped to 85 g/L. However, no blood transfusion was required.

Histopathologic analysis of the biopsy specimen revealed spindle cells, which focally formed irregular vascular spaces; these were in part occluded by thrombi. More solid areas revealed granular iron pigment (Figure 3). Most cells were positive for CD34, which in this setting works as a marker for endothelial differentiation; another endothelial marker, CD31, showed a weak reaction. The morphologic picture was consistent with the aggressive growth of a hemangioendothelioma, most likely of the kaposiform type.
Complete surgical removal was determined to be the best treatment option. Because angiography showed a very highly vascularized tumor with predominant supply from the ethmoid and sphenopalatine arteries, preoperative embolization was performed. The branches of the maxillary artery were successfully embolized, but the ethmoid arteries were not, because of the risk of visual impairment.

To avoid the classical approach of a lateral rhinotomy, a combined approach, entailing an external ethmoidectomy and endoscopic endonasal surgery, was decided upon for the tumor removal. Through a Lynch incision, the tumor was identified in the medial orbit, and the anterior and posterior ethmoid arteries were coagulated bipolarly. Nevertheless, substantial bleeding occurred until the tumor could be freed from the orbital soft tissues and the rhinobasis. Thus, perioperative transfusion was required. Tumor excision was then completed endoscopically. A total sphenoethmoidectomy was performed. The defect in the rhinobasis at the implantation site of the tumor was closed with a mucosal flap elevated from the middle turbinate. Antibiotics (Ceftriaxone) were given at the beginning of surgery and continued until removal of the nasal packing on the fifth postoperative day. The postoperative course was uneventful, and the girl was discharged from the hospital on the sixth postoperative day.

The definitive histologic findings obtained after the removal of the tumor confirmed the suspected diagnosis of a kaposiform hemangioendothelioma. Follow-up consisted of MRI every 6 months for the first 2 years. To date, at 18 months after surgery, the patient is clinically and radiologically free of disease (Figure 2A).

DISCUSSION

The term “hemangioendothelioma” refers to vascular lesions of intermediate or borderline malignancy, showing features between benign heman-
Kaposiform hemangioendothelioma is a locally aggressive vascular neoplasm, characterized by a predominant Kaposi sarcoma-like, fascicular spindle cell growth pattern.\textsuperscript{2–4} Immunohistochemically, these lesions are positive for endothelial antigens, such as CD31, CD34, and factor VIII-related antigen. Focally positive staining for alpha (\(\alpha\)) smooth muscle actin can be noted.\textsuperscript{4}

This tumor occurs most commonly in infants and children, but rare cases have also been described in adults.\textsuperscript{4} Other types of borderline vascular lesions with different histologic and clinical appearances are epithelioid hemangioendothelioma, retiform hemangioendothelioma, polymorphous hemangioendothelioma, and malignant endovascular papillary angioendothelioma (Dabska’s tumor).\textsuperscript{5,6} Spindle cell hemangioendotheliomas, occurring primarily in adults, are now considered benign vascular lesions, for which the term spindle cell hemangioendothelioma was suggested to avoid confusion.\textsuperscript{7,8}

Kaposiform hemangioendothelioma is considered a borderline tumor with a locally aggressive behavior, with no metastatic potential.\textsuperscript{9} An association with lymphangiomatosis has been described for kaposiform hemangioendothelioma, either as diffusely infiltrating lymphangioma or as lymphangioma involving multiple sites.\textsuperscript{3} Some investigators also consider kaposiform hemangioendothelioma benign\textsuperscript{5} or, at least, at the more benign spectrum of borderline tumors. The most common involved sites are cutaneous lesions of the limbs and trunk in 75\% of cases, followed by retroperitoneal involvement in 18\%.\textsuperscript{9} Cases in the head and neck area and the mediastinum have been reported.\textsuperscript{3,10,11}

Kaposiform hemangioendothelioma shows no signs of spontaneous regression. Larger tumors are frequently associated with consumption coagulopathy (Kasabach–Merritt syndrome).\textsuperscript{2–6,9,10} The prognosis depends on the site and size of the lesion. Extensive complete excision seems to be curative. However, the prognosis is poor for patients with large tumors complicated by the Kasabach–Merritt syndrome.

In recent years, several new cases of kaposiform hemangioendothelioma have been reported.\textsuperscript{11,12} This reflects the increased awareness of this rather new tumor entity, with its still widely debated biologic behavior and prognosis. Complete surgical removal is the prime therapeutic option, sometimes resulting in debilitating amputations of extremities.\textsuperscript{3,9} Treatments with prednisone and interferon-\(\alpha\) have been recommended for the Kasabach–Merritt syndrome.\textsuperscript{6,9,10,13}

With regard to the nose and paranasal sinuses, we found no pediatric cases of this distinct tumor described in the English- and German-language medical literature. There exists one similar report of an epithelioid hemangioendothelioma in the nasal cavity of a 23-year-old man.\textsuperscript{14} In contrast to the kaposiform hemangioendothelioma, the epithelioid hemangioendothelioma is considered a low-grade malignant lesion with true metastatic potential.\textsuperscript{5}

This case of a kaposiform hemangioendothelioma arising in a child at an unusual site illustrates the difficulties in making the right diagnosis using imaging techniques and biopsy. In the event of severe epistaxis, this type of tumor should also be taken into account in the differential diagnosis.

\section*{REFERENCES}