OBJECTIVE

To present a rare case of sinonasal non-Ewing’s sarcoma in a pediatric patient. Literature review of sinonasal sarcoma is provided.

BACKGROUND

Sarcomas of the sinuses or nasal cavities are extremely rare in the pediatric population. Tumors in general of the head and neck region are not common in the pediatric population; only about 10% of all pediatric malignancies occur in this location. Of those tumors, even fewer (14%) are malignancies of non-epithelial or connective tissue, and can be characterized as sarcomas. Most sarcomas involving the nasopharynx or paranasal sinuses tend to be large compared to other types of tumors of the head and neck, and are also associated with considerable destruction of bone. Patients present with symptoms that are often nondescript and similar to a common cold. Symptoms can vary from nasal obstruction and nosebleeds to facial deformities, ocular and neurological symptoms in case of large tumors with extra-sinonasal involvement.

Here, we present a report of a sinonasal non-Ewing’s sarcoma in a pediatric patient who presented with facial pain and nasal obstruction. The tumor was initially thought to be an antrochoanal polyp. Complete tumor excision via endonasal endoscopic approach was performed and outcome was monitored by frequent follow-ups.

CASE REPORT

A 10-year-old boy with a history of progressively worsening nasal obstruction and congestion presented with unilateral epistaxis, pain in the nose and maxillary sinuses, and pressure in the left eye. Nasal endoscopy showed a large mass in the left nasal cavity extending toward the maxillary sinus and nasopharynx. Preoperative MRI (Fig. 1) showed an extensive lesion in the left ethmoidal sinus area. Intra-operative images showed a large polypoid mass extending to the maxillary sinus, sphenoid sinus, and partial left frontal sinus. Erosion of the left lamina papyracea was also present.

Complete resection of the tumor was achieved operatively, the patient was discharged one day after surgery, and there is no evidence of recurrent at 27 months after surgery. The patient is currently free of disease, 27 months after surgery (Fig. 3).

DISCUSSION

Accurate diagnosis of this sinonasal sarcoma was made possible by frequent CT scans, which has long been utilized as an optimal radiologic tool to assess and diagnose pediatric craniofacial sarcomas, most notably those arising in the nasal cavity. Sinonasal tumor resection has been correlated with a high incidence of post-surgery survival. However, the location of the tumor may be related to different outcomes – it has been noted that sarcomas in the posterior nasal cavity, nasopharynx, and sphenoid sinus often have worse outcomes than patients with sinonasal sarcomas of other locations. This is primarily due to the difficulty in excising tumors from those specific locations in the nasal cavity. Fortunately the tumor in this case was completely resected using endoscopic sinus surgery.

It is possible that by the time of diagnosis, the sinonasal malignancies may have already started to invade the skull base. About 3% of all brain metastases can be attributed to sarcomas. Due to the physical proximity of sinonasal sarcomas to the skull base, it is recommended to monitor patients routinely for recurrence and to reduce the risk of brain metastases.

CONCLUSIONS

This is a rare case of non-Ewing’s pediatric sinonasal sarcoma fully resected through endoscopic endonasal approach. Patient age and clinical stage of malignancy at initial finding are both correlated with long-term post-diagnosis survival rates.

The patient in this case was young (10 years old), his tumor was diagnosed early and it was low grade. Thus, chemotherapy or radiation was not required and the patient has a good prognosis. Though the patient did develop left frontal sinusitis and required a sinusotomy one year after tumor excision, nasal function was eventually restored. The patient is currently free of disease, 27 months after surgery.

Figure 1. Pre operative T1 weighted MRI of the orbit with contrast: (A), (B) Axial views. (C), (D) Coronal views. Note the mass effect is present on the left lamina papyracea without intracranial invasion. The tumor extends to the maxillary sinus and it is in close relation to the skull base.

Figure 2. Endoscopic intraoperative view with 0 degree endoscope (A, B and C) and 45 degree endoscope (D). (A) Tumor is seen between the inferior turbinate and septum. (B) Tumor extending to the left lamina papyracea without intraconal invasion. (C) Cavity after the resection. (D) Frontal sinus.

Figure 3. Post operative T1 weighted MRI: (A), (B) Axial views. (C), (D) Coronal views. Sinuses are open and there is no evidence of recurrent at 27-months after the surgery.