To report a rare case of hereditary hemorrhagic telangiectasia and olfactory schwannoma completely resected with a unilateral extended endoscopic endonasal approach.

Olfactory schwannomas of the anterior skull base (ASB) are rare tumors and their association with hereditary hemorrhagic telangiectasia (HHT) has not previously been described.

The majority of ASB schwannomas arise from the sinonasal tracts, and may demonstrate intracranial extension. We report a case with an olfactory schwannoma of the ASB with subpial involvement of the frontal lobe. This is the first reported single stage fully endoscopic endonasal unilateral approach for resection of an olfactory schwannoma with preservation of the contralateral olfactory cleft.

A twenty-five–year–old female with a history of migraines and hereditary hemorrhagic telangiectasia (HHT) presented with unilateral episodic epistaxis and progressive worsening of her headaches. She denied any symptoms of nasal obstruction or recurrent sinusitis. Nasal endoscopy showed mild telangiectasias in the mucosa bilaterally, but no clear evidence of a mass. The middle meatus anatomy on the left side was preserved. A CT scan showed an expansive lesion in the left ethmoid area with bone remodeling, suggesting a longstanding benign tumor, measuring 3 cm antero-posterior, 2.7 cm transverse, and 3.2 cm craniocaudal (Figure 1). Subsequent MRI with contrast showed the mass to be avidly enhancing with intratumoral flow voids (Figures 2A, 2B and 2C). Definitive radiographic evidence of brain invasion was not present, although there was mass effect and displacement of the overlying frontal lobe and the adjacent left orbit. Due to the significant contrast enhancement in T2WI and patient with HHT, catheter angiography was performed, revealing tumor supply largely from the left anterior ethmoidal artery with a mild vascular blush. The intracranial anterior circulation was displaced superiorly and posteriorly by the mass.

The patient underwent unilateral anterior cranial base resection via an extended endoscopic endonasal approach (Figure 2). Despite not appearing overly vascular on the preoperative angiogram, the ipsilateral sphenopalatine artery (SPA) was ligated to decrease the blood flow to the nasal cavity due to the patient's history of HHT. The ethmoidal arteries were cauterized with bipolar as usual step in endoscopic anterior cranial base resections. The tumor margins were identified, with a dissected plane laterally along the orbit and periosteum as well as medially along the nasal septum. After sufficient internal debulking of the tumor was performed, sharp extracapsular dissection was utilized. The mass was largely contiguous with the left anterior cranial base dura, necessitating a circumferential dural resection. Superiorly, the arachnoid adhesions to the basal frontal lobe were sharply divided. In the region of the gyrus rectus, dense pial adherence was noted but extracapsular dissection was able to performed. The olfactory nerve and bulk were seen emerging from the posterior aspect of the tumor and were sharply ligated proximally. The tumor remnants and its dural attachment were then completely removed through the left unilateral approach. Dural and olfactory nerve margins were resected and sent for intraoperative pathologic examination, demonstrating no evidence of residual tumor (Figure 4). The left anterior cranial base dura was primarily repaired with an inlay collagen synthetic graft. Contralateral nasoseptal flap was used due to the ligation of the left SPA. The flap was transposed to the left side through a partial posterior septectomy.

Ten months postoperatively, the patient continues to do well and nasal endoscopy demonstrates satisfactory sinonasal healing. The patient's sense of smell was preserved, based on the University of Pennsylvania Smell Identification Test (UPSIT®). Follow-up MRI shows no evidence of tumor residual or recurrence at this early timepoint (Figures 2D, 2E and 2F).

We report a rare case of HHT and olfactory schwannoma completely resected with a unilateral extended endoscopic endonasal approach. Reconstruction was performed with a contralateral nasoseptal flap. This is the first reported single stage fully endoscopic endonasal unilateral approach for resection of an olfactory schwannoma with preservation of the contralateral olfactory cleft. The sense of smell and taste is preserved postoperatively.