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First Description and Surgical Treatment of a Primary Frontobasal Ganglioneuroma: Case Report

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ABSTRACT

Background: Ganglioneuromas are rare, benign tumors arising from the sympathetic or parasympathetic nervous system, most commonly located in the mediastinum, retroperitoneum, or adrenal glands. Skull base involvement is extremely rare, and frontobasal ganglioneuromas have not been previously reported.

Case Presentation: We report the case of a 52-year-old women presenting with watery rhinorrhea, anosmia, nasal obstruction, intermittent visual disturbances, and refractory vascular hypertension. Imaging revealed a frontobasal mass extending into the nasal cavity. The patient underwent endonasal endoscopic resection under neuronavigation guidance, including partial resection of the nasal septum. Dural reconstruction was performed using a multilayer underlay-and-overlay technique with autologous fascia lata.Complete tumor resection was achieved without intra- or postoperative complications. Postoperative follow-up demonstrated satisfactory mucosal healing, no cerebrospinal fluid leakage, and no evidence of local recurrence. Histopathological analysis confirmed a ganglioneuroma.

Conclusion: This is the first reported case of a primary frontobasal ganglioneuroma. Endonasal, navigation-guided resection allowed complete tumor removal with favorable functional and reconstructive outcomes. Ganglioneuroma should be considered in the differential diagnosis of primary frontobasal masses, despite its rarity.

Keywords: *Surgical Treatment, Primary Frontobasal Ganglioneuroma, Benign Tumors*

Introduction

Ganglioneuromas are rare, benign tumors deriving from neural crest cells and composed of mature ganglion cells and Schwann-type elements. They are part of the spectrum of neuroblastic tumors. Unlike neuroblastomas or ganglioneuroblastomas they are well-differentiated and slow-growing. Common anatomical sites include the posterior mediastinum, retroperitoneum, adrenal glands, and cervical sympathetic chain (Shah *et al*., 2021; Kayastha *et al*., 2020). Clinical symptoms are often related to mass effect rather than biochemical activity (Yousefi *et al*., 2023).

Intracranial ganglioneuromas are exceedingly uncommon. A number of reports describe ganglioneuromas involving cranial nerves (especially the trigeminal nerve) and skull base regions, with varied clinical presentations such as trigeminal neuralgia, headache, and facial numbness (Kim *et al*., 2013). Another report described a ganglioneuroma of the internal auditory canal, expanding the spectrum of skull-base involvement (Ozluoglu *et al*., 2007). Additionally, ganglioneuroma involving the skull base bones, such as a lesion of the sphenoid wing, has been reported, though even these tend to be intraosseous rather than arising from the frontobasal dura or region (Aktüre *et al*., 2011).

To date, no published case has described a primary ganglioneuroma located at the frontobasis in an adult, causing symptoms such as nasal obstruction, rhinorrhea, anosmia, visual disturbance, or refractory hypertension. The anatomical and functional complexity of the frontobasal region—including proximity to the nasal cavity, sinuses, optic apparatus, and anterior cranial fossa dura—raises particular surgical challenges.

Herein we present a case of a 52-year-old patient with a frontobasal ganglioneuroma, who underwent complete endonasal endoscopic resection under navigation, with dural reconstruction, histopathological confirmation, and imaging follow-up. This case contributes a novel anatomical location to the literature, underscores the diagnostic challenges in frontobasal masses, and demonstrates that a minimally invasive endonasal approach can be both effective and safe in this setting.

Case Presentation

A 52-year-old women presented with a six-month history of watery rhinorrhea, progressive nasal obstruction, anosmia, and intermittent visual disturbances. Additionally, the patient suffered from severe vascular hypertension that remained refractory to triple-combination antihypertensive therapy. There was no prior history of trauma, surgery, or sinonasal disease.

On clinical examination, anterior rhinoscopy and nasal endoscopy revealed a mass occupying the superior nasal cavity with contact to the nasal septum and skull base. Neurological examination was unremarkable apart from anosmia. Magnetic resonance imaging demonstrated a well-circumscribed, frontobasal mass extending into the nasal cavity, showing homogeneous contrast enhancement with signs of infiltration into surrounding structures. Computed tomography showed partial destruction adjacent bony structures (Fig. 1). Differential diagnoses included meningioma, olfactory neuroblastoma, and schwannoma.

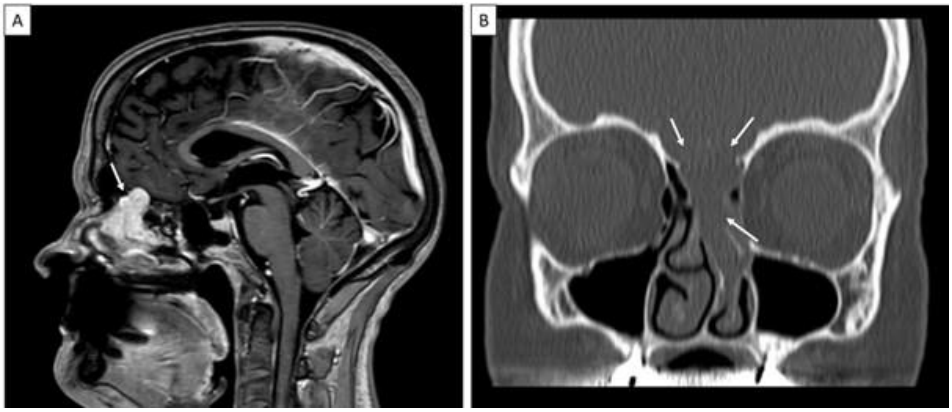

Figure 1: (A) Sagittal T1-weighted contrast-enhanced MRI demonstrating a well-circumscribed frontobasal lesion with predominantly homogeneous enhancement, contacting the inferior frontal lobe and extending into the nasal cavity. (B) Coronal MRI view of the centrofacial region revealing partial destruction of adjacent bony structures, including erosion of the central skull base and superior nasal septum. Arrows indicate tumor borders.

The surgical intervention was performed via an endonasal endoscopic approach under neuronavigation guidance. After careful exposure of the frontobasal mass, the tumor was dissected from surrounding structures and completely removed including resection of the superior septum, parts of the skull base and the infiltrated dura (Fig. 2B). During the procedure, complete tumor resection resulted in an extended dural defect at the anterior skull base. For watertight closure and to prevent postoperative cerebrospinal fluid leakage, a multilayer reconstruction was performed using autologous fascia lata harvested from the right thigh. The graft was placed in both underlay and overlay fashion to reinforce the dural repair. Hemostatic agents, including Tabotamp® and TachoSil®, were applied to ensure additional sealing and promote tissue adhesion and were finally covered with a free mucosal flap from the left middle turbinate (Fig. 2C-E). After reconstruction, no visible liquorrhea or CSF excretion was observed endoscopically. The surgical field was dry, and the nasal cavity was gently packed for stabilization of the reconstruction.

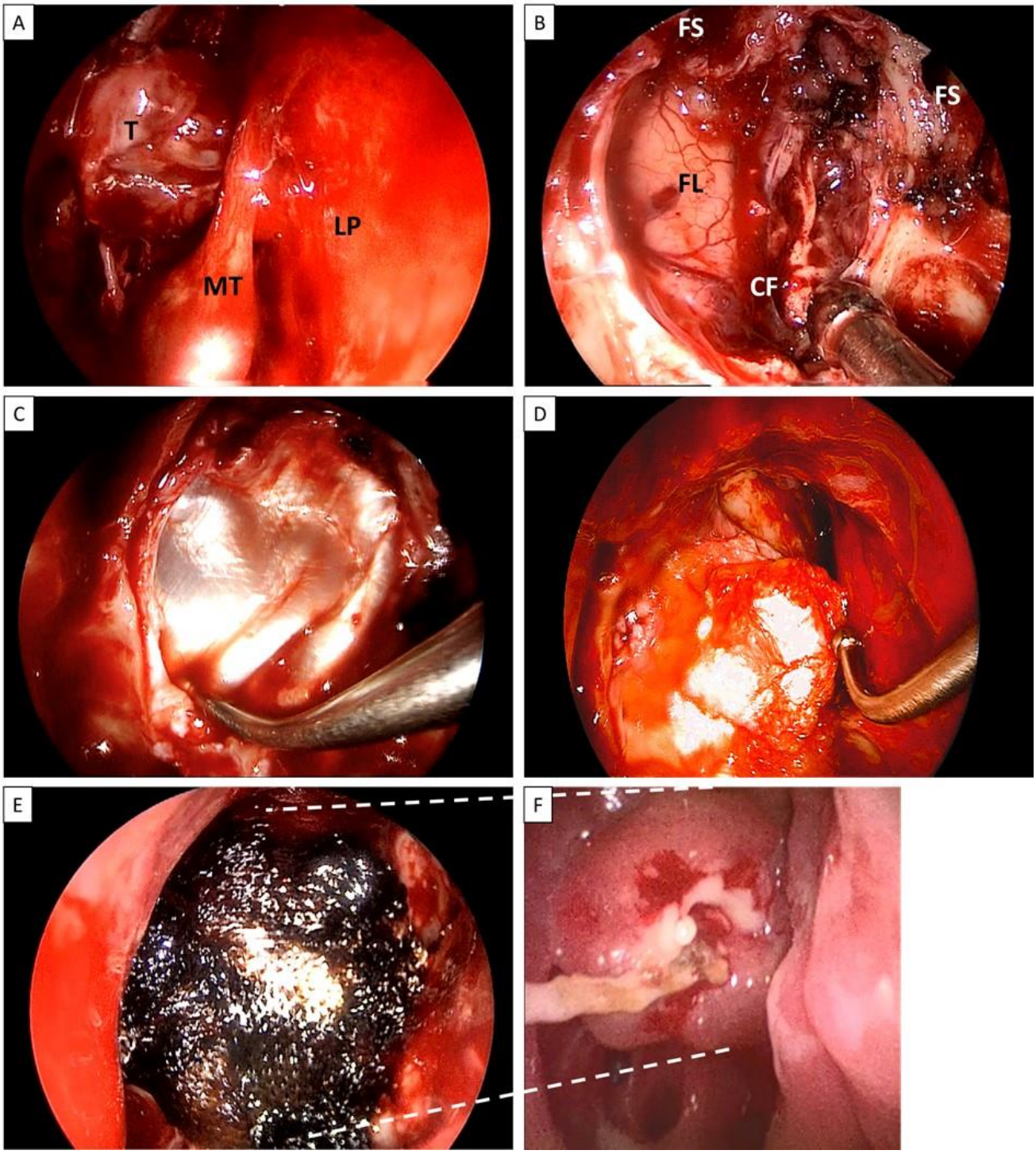

Figure 2: (A) Endoscopic intraoperative view showing the frontobasal tumor (T) adjacent to the middle turbinate (MT) and lamina papyracea (LP). (B) Post-resection view of the surgical field illustrating the exposed frontal sinus (FS), frontal lobe (FL), and cerebral falx (CF) following complete endoscopic tumor removal. (C–E) Sequential images demonstrating reconstruction of the skull base defect using an autologous fascia lata graft, reinforced with Tabotamp® and TachoSil®. The hemostatic materials turned dark upon contact with blood, confirming activation. (F) Endoscopic follow-up examination 8 weeks postoperatively showing complete mucosal coverage with minimal central scarring and no evidence of inflammation or cerebrospinal fluid leakage.

Postoperatively, nasal packing was left in place for five days to provide mechanical support and counteract caudal pressure on the duraplasty. The patient received systemic antibiotic prophylaxis with ceftriaxone 2 g intravenously once daily for five days. Close clinical follow-up was performed during the postoperative period to monitor for potential complications such as infection, CSF leakage, or graft displacement.

The postoperative course was uneventful. The patient showed rapid improvement of nasal breathing and resolution of rhinorrhea. Histopathological and immunohistochemical analysis of the specimen confirmed the diagnosis of a mature ganglioneuroma, characterized by mature ganglion cells embedded within a Schwannian stroma, positive for S100 (Fig. 3). Follow-up MRI and nasal endoscopy at three and six months demonstrated complete resection, satisfactory mucosal healing, and no evidence of recurrence or dural defect (Fig. 2F).

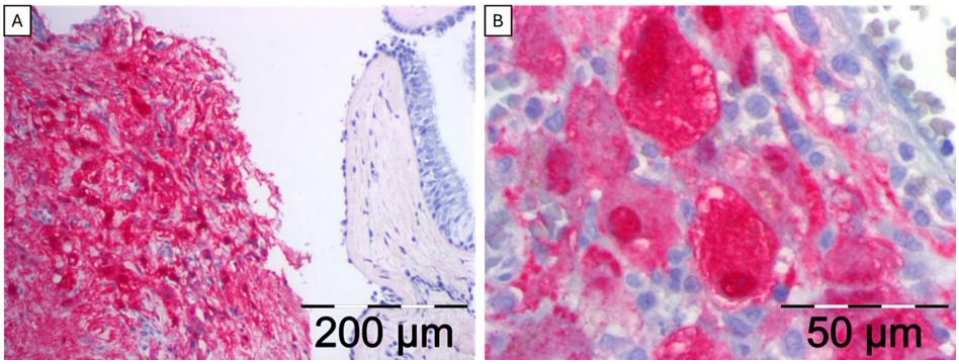

Figure 3: Histopathological and immunohistochemical findings of the resected tumor: (A) The lesion is in close proximity to ciliated respiratory epithelium, indicating juxtaposition to normal epithelial structures. (B) Immunohistochemical staining for S100 highlights the Schwannian stroma of the ganglioneuroma, showing intense cytoplasmic and nuclear positivity.

Discussion

Ganglioneuromas are rare, benign tumors originating from neural crest cells, typically arising from sympathetic ganglia. While most commonly found in the posterior mediastinum (60–80%), retroperitoneum, and adrenal glands (10-15%), their occurrence in the skull base is exceedingly rare (Gasparini *et al*., 2024). This case report highlights the diagnostic and therapeutic challenges associated with such an uncommon presentation.

The patient presented with a constellation of symptoms, including watery rhinorrhea, anosmia, nasal obstruction, intermittent visual disturbances, and refractory hypertension. These nonspecific symptoms posed a diagnostic challenge, as they overlap with various sinonasal and intracranial

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pathologies. In previously reported head and neck ganglioneuromas, symptoms typically relate to mass effect—most commonly nasal obstruction, epistaxis, or visual disturbances, depending on location. In cases where they exhibit low neuroendocrine activity, they may lead to symptoms such as diarrhea, hypertension, virilization, and myasthenia gravis (Gasparini *et al*., 2024).

Imaging studies revealed a well-circumscribed, homogeneously enhancing mass at the anterior skull base, extending into the nasal cavity, and eroding adjacent bony structures, including the superior nasal septum and central skull base. Such imaging characteristics are consistent with previously reported cases of ganglioneuromas located in the head and neck region (Ma *et al*., 2012).

A brief review of the literature shows only isolated instances of central or skull-base ganglioneuromas, including lesions of the trigeminal nerve, and parapharyngeal space and larynx (Gasparini *et al*., 2024; Ma *et al*., 2012; Narra *et al*., 2024)—none of which involved a primary frontobasal origin. Compared with these cases, the present lesion demonstrated a more pronounced anterior and inferior extension into the nasal cavity, contributing to the patient's sinonasal symptomatology.

Given the tumor's location and the patient's symptoms, an endonasal endoscopic approach was appropriate. This minimally invasive technique allowed complete resection of the tumor while preserving adjacent structures. Recent advances in endoscopic anterior skull base surgery—demonstrating high rates of complete resection with low morbidity and reliable multilayer reconstruction outcomes—support the use of a minimally invasive endonasal approach for anterior skull base lesions such as in our case (Kikuchi and Nakagawa, 2023; Abiri *et al*., 2020). This time dural reconstruction was achieved using autologous fascia lata in a multilayer underlay-and-overlay technique, reinforced with hemostatic agents Tabotamp and TachoSil and covered by a free mucosal flap. Postoperative imaging confirmed complete resection with no evidence of cerebrospinal fluid leakage or recurrence. Although complete resection is generally curative and recurrence is considered rare, the true long-term recurrence risk is not fully established due to the scarcity of cases and limited follow-up in the literature

While ganglioneuromas are predominantly peripheral lesions, their occurrence in the central nervous system is exceedingly rare. Reported cases have included tumors arising from cranial nerves, such as the trigeminal nerve (Narra *et al*., 2024), and other regions within the head and neck (Ma *et al*., 2012).

This case highlights the diagnostic complexity and surgical considerations associated with ganglioneuromas in uncommon locations. Given their rarity, ganglioneuromas should be included in the

differential diagnosis when evaluating enhancing masses in the anterior skull base. Early recognition and appropriate surgical intervention can lead to favorable outcomes, as demonstrated in this patient.

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