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Case Report

Navigation-assisted endoscopic endonasal surgery of a glomangiopericytoma with intraorbital extension: A case report and literature review

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Abstract

A glomangiopericytoma, or sinonasal type hemangiopericytoma, is a rare lesion which accounts for <0.5% of all sinonasal tumors. The mainstay treatment is wide excision. Instead of traditional open surgical approaches, such as midfacial degloving or lateral rhinotomy, we offer a case of 21-year-old male with diagnosis of glomangiopericytoma with skull base and intraorbital invasion and received navigation-assisted endoscopic excision of a glomangiopericytoma.

Keywords: *Endoscopic surgery, Glomangiopericytoma, Hemangiopericytoma, Navigation*

Introduction

The mainstay treatment is wide surgical excision with clear margins. In the past, patients with intraorbital or skull base invasion usually received open craniofacial resection, midfacial degloving, or lateral rhinotomy, all of which were associated with significant morbidity. However, recent advances in endoscopic surgery with navigation guidance have achieved fair surgical results with less morbidity than the traditional methods [3-5]. Here, we report the excision of a glomangiopericytoma with skull base and intraorbital invasion using navigated endoscopic surgery. We also describe the features and possible therapeutic options for glomangiopericytomas based on a literature review.

Case report

A 21-year-old male student with no known underlying diseases was admitted through the emergency department for intermittent epistaxis for 3 months. He also had nasal obstruction and anosmia. Ophthalmologic symptoms, including diplopia and proptosis, started 1 month before admission. Local findings revealed a unilateral polypoid lesion obscuring the left osteomeatal complex with active bleeding. Nasal pledget packing could not stop the bleeding. Emergency endoscopic surgery for hemostasis and biopsy was done. The pathology revealed a glomangiopericytoma.

Computed tomography showed a mass lesion occupying the left maxillary sinus, middle meatus, ethmoid sinus, frontal recess, and frontal sinus. Obvious mass effect with the surrounding structure deviation was observed, and bone destruction was highly suspected [Figure 1]. The left lamina papyracea and cribriform plate could not be identified in the preoperative image. Due to the clinical symptoms, left orbital cavity and anterior skull base tumor invasion were impressed. Subsequent angiography revealed a hypervascular tumor supplied predominantly by the left internal maxillary artery, and embolization was performed preoperatively.

The Fusion™ ENT Navigation System and powered ENT instruments (Medtronic, Mounds View, MN, United States) were applied because of severe distortion of the anatomy. The procedures were performed under general anesthesia. After

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decongesting the nasal mucosa with adrenaline-soaked cotton pledgets, the nasal cavity was inspected with a 0° endoscope first. The left nasal middle meatus was full of tumor and landmarks could barely be identified. Under navigational guidance, tumor debulking was performed and we identified the tumor pedicle at the frontal recess. A wide endoscopic resection of the surrounding tissue was performed through a maxillary sinus antrostomy, turbinectomy, complete sphenoethmoidectomy, and frontal sinusotomy. At least a 3–5 mm margin was left. For the tumor pedicle at the frontal recess, a drill was applied to remove the tumor and bone chip underneath as a deep margin. The dura was exposed during the procedure and was well preserved. The cribriform plate was deformed but not involved by the tumor. For the orbital cavity lesion, the lamina papyracea was deformed, and it was removed and we found that the periorbita was free from invasion. Frozen section analysis of the bony margin of the frontal recess and lamina papyracea was performed, and the margins were free from tumor invasion.

The final pathology revealed a low-grade glomangiopericytoma [Figures 2 and 3]. The postoperative course was uneventful, and the patient remained disease free during 12 months of follow-up [Figure 4].

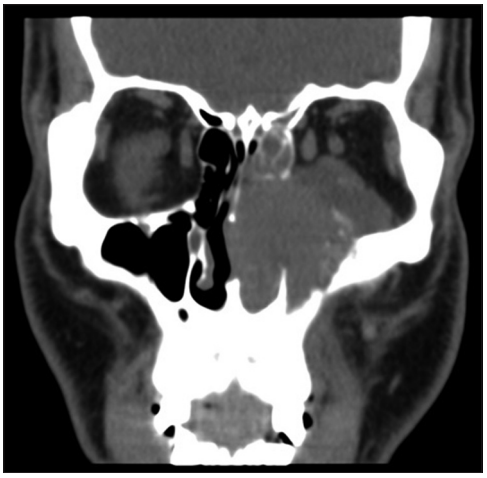

Figure 1: Preoperative computed tomography scan. A mass lesion occupies the left maxillary sinus, nasal cavity, and ethmoid sinus with bone destruction. The nasal septum is deviated to the right due to mass effect

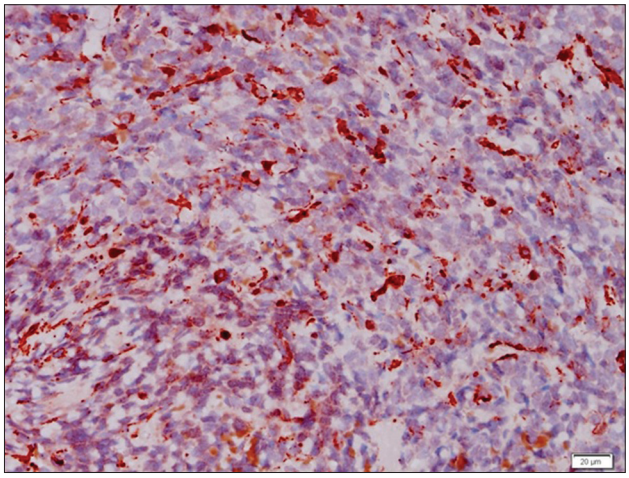

Figure 3: Positive anti-vimentin stain

Discussion

A glomangiopericytoma is a very rare lesion and the most common presenting symptoms are nasal obstruction and epistaxis. With extension into the orbital cavity, diplopia and proptosis may be observed. Surgical excision is still the mainstay treatment. Clinically, glomangiopericytomas are more indolent than hemangiopericytomas that occur elsewhere in the body. However, glomangiopericytomas are histologically malignant tumors with metastatic potential, and the reported recurrence rate for incomplete resection ranges from 7% to 50%. The average time to recurrence is 6–7 years [6]. In the past, open surgery, such as craniofacial resection, lateral rhinotomy, or midfacial degloving, was the only strategy to achieve free margins. Based on the literature [3-5,7-12], the endoscopic approach has gained acceptance as long-term results have become available and has demonstrated comparable

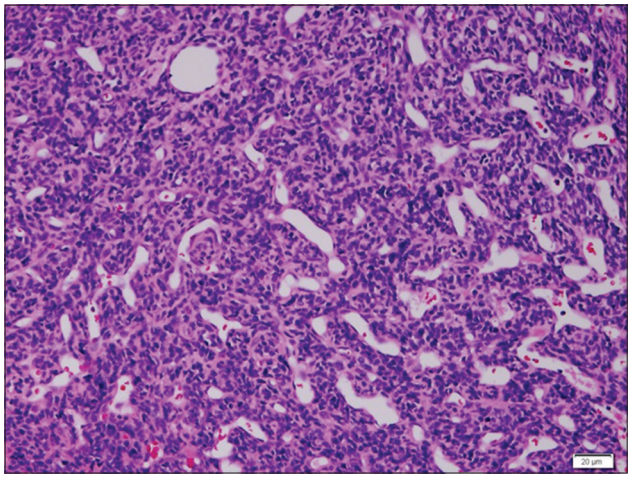

Figure 2: Microscopically, the tumor is comprised of closely packed cells, forming short fascicles and focally exhibiting a storiform, or palisaded pattern, interspersed with many "staghorn-like" vascular channels. The tumor cells are uniform and elongated-to-oval shaped with round-to-oval-to-spindle-shaped nuclei. There are mild nuclear pleomorphism and a little mitotic activity, with <4/10 per high-power field. Therefore, this sample represents a low-grade glomangiopericytoma

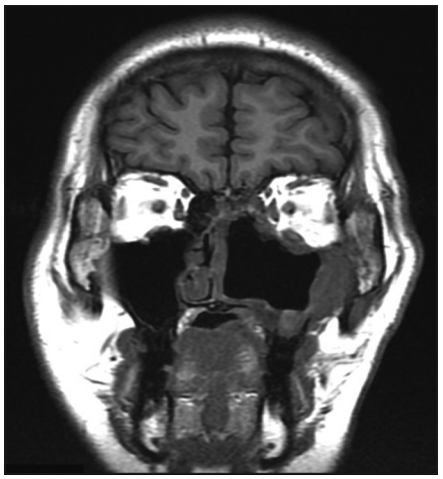

Figure 4: No evidence of residual tumor at 12 months postoperatively

Table 1: Literature review of glomangiopericytoma patients undergoing endoscopic resection

Authors	Patient number	Location	Complications	Mean follow-up (months)	Local recurrence	Time to recurrence (months)
Bhattacharyya *et al*. [12]	1	Left posterior ethmoid cells	0	24	0	-
Serrano *et al*. [11]	5	2 in left nasopharynx and ethmoid cells 1 in right posterior ethmoid cells and sphenoid sinus (recurrence) 1 in left posterior ethmoid cells 1 in right posterior ethmoid cells and sphenoid sinus	0	54	1	60
Castelnuovo *et al*. [10]	1	Right nostril with cribriform plate invasion	1 (predicted CSF leak)	48	0	-
Poetker *et al*. [5]	1	Middle turbinate	Not mentioned	13	0	-
Bignami *et al*. [4]	10	Not mentioned	1 (stroke)	42.5	1	73
Arpaci *et al*. [9]	1	Left nostril with extension into nasopharynx	0	Not mentioned	0	-
Gomez-Rivera *et al*. [8]	6	Not mentioned	2 (bleeding and pneumonia)	25	0	-
Tessema *et al*. [3]	12	Not clearly mentioned (6 involved cribriform plate; 1 frontal recess; 3 sphenoid sinus)	0	41	0	-
Sun *et al*. (2013, this patient)	1	Left maxillary sinus, left nostril, and ethmoid sinus with extension into left orbital cavity and anterior skull base	0	12	0	-

CSF: Cerebrospinal fluid

disease-free survival and recurrence rates with excellent functional outcomes and a better quality of life [Table 1]. Major complications such as postoperative cerebrospinal fluid leak, meningitis, massive epistaxis requiring blood transfusion, stroke, and death have rarely been reported or could be predicted preoperatively. Other treatment modalities such as adjuvant radiotherapy or chemotherapy are still controversial, and the benefits have not been ascertained [13]. However, in patients with a positive margin, adjuvant therapy could be considered. In conclusion, the current data show equivalent outcomes for open and endoscopic surgery and less morbidity for the endoscopic method; thus, endoscopic excision should be considered first.

Declaration of patient consent

The authors certify that the patient has obtained appropriate patient consent form. In the form, the patient has given his consent for images and other clinical information to be reported in the journal. The patient understands that his name and initial will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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