

Malignant Nasosinusal Glomic Tumor. A Rare Case Presentation and Literature Review

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2. Keywords

Glomus tumors; Nasal location; Pathological findings

1. Summary

Glomus tumors are lesions that arising from the glomus neuromyoarterial body, their location in the head and neck is rare.

We present a case of a nasosinusal glomus tumor in a 70-year-old female patient with right nasal respiratory failure and right otic plugging, initially diagnosed as nasosinusal polyposis and treated surgically with endoscopic sinonasal surgery.

Findings of pathological anatomy of difficult histological characterization revealed right nostril nasosinusal glomus tumor and left nostril malignant tumor. Due to unexpected pathological results, we performed excision by endoscopic sinonasal surgery and adjuvant radiotherapy.

3. Introduction

The glomus is a thermoregulatory shunt composed of a neuromyoarterial body, consisting of an afferent artery, an efferent venula, and multiple communications between them. This arteriovenous anastomosis is surrounded by glomal cells, which have a contractile capacity and provide the glomal body with the ability to contract or relax them when faced with thermal or pressure stimuli resulting in regulation of the vessel's caliber and the blood flow.

Glomus tumors are mostly benign lesions. Most frequent location is the distal part of the extremities, specifically the subungual bed [1]. However, they have also been described in other locations, although very infrequently in a nasosinusal location, as in the case described in this article. This tumor entity is difficult to characterize histologically. It is believed to derive from the muscle and endothelial cells of the glomus, but its definitive immunohistochemical characterization has yet to be defined.

4. Clinical Case

A 70-year-old woman came to our service with a 10 months history of right nasal respiratory failure and right otic plugging. On physical examination, otoscopy was normal, rhinoscopy and endoscopy showed pale right inferior turbinate and bilateral polypoid tissue from the superior meatus.

Facial CT scan study evidenced soft tissue located in the right nasal fossae, occupying almost all of it, with a diameter of 47 mm. In addition, CT scan showed a second lesion in the left nasal fossae, with a diameter of 25 mm. Retention of secretions in ethmoid cells and right portion of the sphenoid sinus. It is concluded as bilateral

nasosinusal polyposis, with predominance in right side (Figure 1).



Figure 1: Facial CT

With these findings, the patient was treated by Endoscopic Nasosinusal Surgery. Intraoperative findings were bilateral polyposis from the superior meatus, grade IV in the right nostril and grade II in the left one. Middle meatus was free of polyposis.

Histopathological study showed a densely cellular spindle cell lesion, made up of monotonous, oval-nucleus cells arranged in irregularly crossed longitudinal bundles.

Immunohistochemically the lesion was positive for Factor XII, CD34 and focally for smooth muscle actin. Stains for collagen IV, S100, EMA, desmin, H-Caldesmon and STAT6 was negative. The FISH technique to assess SYT rearrangement was negative. Submucosal lesion formed by a proliferation of poorly defined eosinophilic cytoplasm epithelioid cells and monotonous, round nuclei without atypia. Rich vascularity with hemangiopericytoid pattern.

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Immunohistochemically there is diffuse positivity for smooth muscle actin and focal positivity for collagen IV.

It was concluded a fusocellular tumor that presents as a first diagnostic option a malignant glomus tumor.

Post-operative MRI of the head and neck showed occupation of both maxillary sinuses by secretions. Also it showed a soft tissue thickening at the ethmoid level, in the bilateral posterior medial region, intensely enhanced after the administration of intravenous contrast and suggestive of tumor remains (Figure 2). No other morphological or signal alterations were observed at the level of the laryngeal pharynx structures and oral cavity, nor pathological lymphadenopathy in the different neck ganglion chains. (Figure 2)



Figure 2: MRI Head and neck

Angio CT scan revealed a soft tissue with tumoral features, with a difference in the appearance of the posterior and medial mucous secretions of the sinus cells. Also CT detected a bilateral ethmoidal image extended to the anterior margin of the homolateral sphenoid on the right side, with intense accumulation of the venous phase (Figure 3).

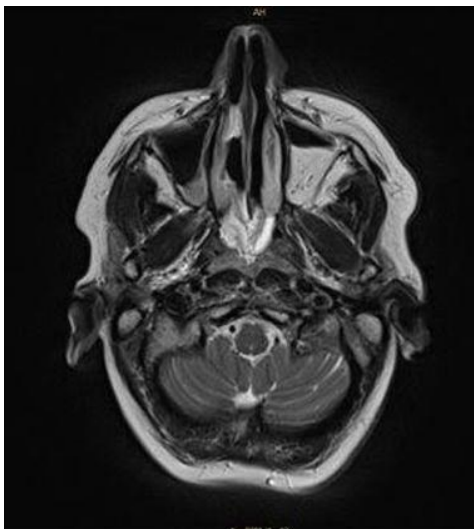


Figure 3: Angio CT

After 3 months of the first evaluation, the patient underwent surgery for exeresis and resection of a malignant glomus tumor by nasal endoscopy with a neuronavigator. A right partial mid turbinectomy was performed to improve the approach to the area of residual lesion. Polypoid tissue was seen into the ipsilateral sphenoid sinus, and occupied areas of the posterior ethmoid.

An ethmoidectomy of the entire posterior and medial area was performed, until the pathological appearance of the mucosa was no longer visible.

Histological study of sample tissue showed glomus tumor with reactive changes without evidence of malignance.

Control head and neck MRI after 6 months of the first surgical intervention evidenced right middle turbinectomy image with bilateral endoscopic ethmoidectomy. Furthermore MRI showed a diffuse inflammatory thickening of the mucosa surface of the endonasal lining at the bilateral ethmoid level and the lining mucosa of both maxillary sinuses. In morphological sequences, a clear tumor residue is not more identified. However, in the dynamic post-contrast enhancement sequence, an early laminar enhancement (2mm thick) at level of the left ethmoid ceiling might suggest the presence of a small lamellar tumor debris at this level (Figure 4).

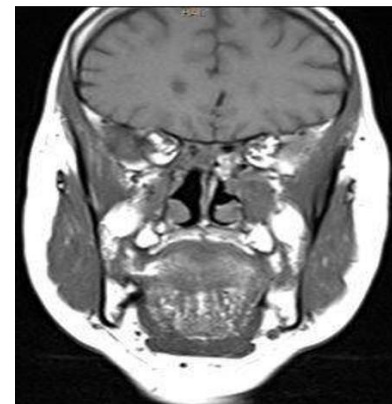


Figure 4: MRI Head and neck

Because these findings, the patient received 33 sessions of 66 Gy for 2 months. In the MRI control study at 9 months, no images suggesting tumor remains were found.

5. Discussion

Glomus tumors are benign lesions derived from the glomus, a neuro-arterial structure that helps regulate body temperature and are found predominantly in the distal areas of the body [2]. They have also been described in other more atypical locations such as bone, trachea, lung, gastrointestinal tract, and head and neck. Glomus tumors of the sinonasal tract are very rare, less than 0.5% of nasosinusal non-epithelial tumors [3, 4, 7]. In fact, less than thirty cases have been reported in the literature until 2020. Within the nasosinusal tract, the most frequently described location has been

the nasal septum [2].

We present a 70-years-old patient who consulted for a ten-month-old right nasal obstruction. The age of our patient is higher than the average found in the literatura. We also observed a prevalence of the female gender with respect to the male gender of 2: 1 [2, 4, 5].

The symptoms most frequently described in the literatura are nasal obstruction, recurrent epistaxis, rhinorrhea, persistent facial pain. The glomus tumors tend to be described macroscopically as a single, generally red-grayish, rigid mass of nodular or polypoid morphology. In our case, due to the morphological appearance, the first differential diagnosis was the polypoid inflammatory lesion of both nostrils.

The classic histological presentation of a glomus tumor is a row of small round monomorphic perivascular cells. Their nuclei are oval and poor in chromatin, with an eosinophilic cytoplasm. Normally they are arranged on a stroma that may have hyaline degeneration, mucoid or have hemosiderin deposits as a consequence of possible trauma. In the literature, three histological subtypes of the classic glomus tumor are described: the glomangioma, glomangiomyoma, and the solid glomus tumor. These subdivisions are made according to the majority component.

The immunohistochemistry of all these lesions (an ingrained positivity for the MSA and α -SMA markers) suggests the origin of glomus tumors as transitional cells between muscle and endothelial cells [5]. The pathological anatomy of the lesions in our case coincides with the review in the literatura was presented in the left nostril as epithelioid cells of poorly defined eosinophilic cytoplasm with monotonous, oval nuclei and without atypia. On the other hand, an infrequent finding where a loss of differentiation is verified, a clear relationship with the sample of the right nostril persisting (clearly characterized as a classic glomus tumor). Immunohistochemistry was also different between left nostril (possible malignancy glomus tumor) positive for factor XII, CD34 and focally for smooth muscle actin, compared with contralateral lesion (glomus tumor), with diffuse positivity for smooth muscle actin and collagen IV. The CD34 staining was negative. A more widespread CD34 pattern and not just circumscribed to pseudocapsular epithelial cells, it can lead to an infiltrative glomus tumor.

Treatment is total excision of the lesion [3, 4, 6]. In the presented case, due the differentiation found in the left nostril lesion and the findings of postoperative imaging studies, we decide in the ENT oncology committee, to perform an adjuvant treatment with radiotherapy.

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