Glomangiopericytoma of the nasal cavity: A case report

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ABSTRACT

Introduction: Glomangiopericytoma is a rare perivascular tumor that accounts for less than 1% of all nasosinusal tumors. It belongs to the category of low malignancy tumor with a good prognosis.

Case report: We describe a case of 48-year-old female presented to our department with 2 months' history of progressive, permanent left nasal obstruction and nasal bleeding. Endoscopic examination of the nasal cavity revealed a left-sided, friable, vascular mass abutting to the superior turbinate. Computed tomography (CT) scan and Magnetic resonance imaging (MRI) showed a soft tissue mass occupying the left nasal cavity without bony destruction with intense contrast enhancement on MRI. The tumor was treated by endoscopic resection. Histological and immunohistochemical examination retained the diagnosis of glomangiopericytomas. During the 12-months follow-up no recurrence was observed

Conclusion: Glomangiopericytomas are rare, vascular tumors. It generally arises in the nasal cavity and may extend into the paranasal sinuses. Successful management depends on complete resection.

Key words: Glomangiopericytoma, Sinonasal cavity, Endoscopic surgery.

INTRODUCTION

Glomangiopericytoma (GPC) is a rare sinonasal mesenchymal neoplasm arising from the pericytes surrounding the capillaries. It accounts less than 0.5–1% of all sinonasal tumors [1]. It was first reported as a hemangiopericytoma in 1942 by Stout and Murray; however, in 2005 the World Health Organization (WHO) classified this disease as GPC. It was defined as a sinonasal neoplasm demonstrating a perivascular myoid phenotype [2]. It belongs to the category of borderline and low-malignant-potential soft tissue tumors of the sinonasal tract [2]. Our aim was to discuss diagnosis and therapeutic approaches of this rare tumour.

CASE REPORT

A 48-year-old female presented to the ENT Department of Farhat Hached Hospital with 2-month history of progressive, permanent left nasal obstruction and nasal bleeding. She had no anosmia no ophthalmologic symptoms and no headache. Endoscopic examination of the nasal cavity revealed a left-sided, friable, vascular, nasal mass abutting the superior turbinate. The floor and the nasal septum were not involved. There was no lymphadenopathy on the cervical palpation.

A computed tomography (CT) scan showed a soft tissue mass of 3 cm x 1.5cm occupying the left nasal cavity with a heterogeneously enhancing. There was no bony destruction or invasion of the surrounding tissue (Figure 1).

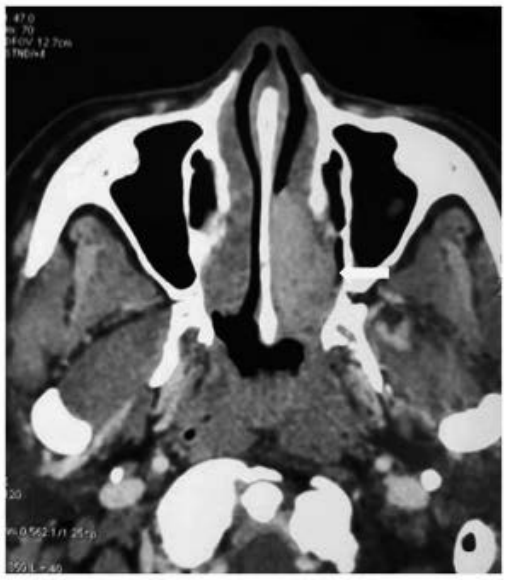

Figure 1 : Axial enhanced CT scan showing soft tissue density in the left nasal cavity with heterogeneous enhancement with no involvement of the maxillary sinus.

Magnetic resonance imaging (MRI) showed a polypoid mass isointense on T1-weighted, hyperintense on T2-weighted with intense contrast enhancement on the left nasal cavity (Figure 2).

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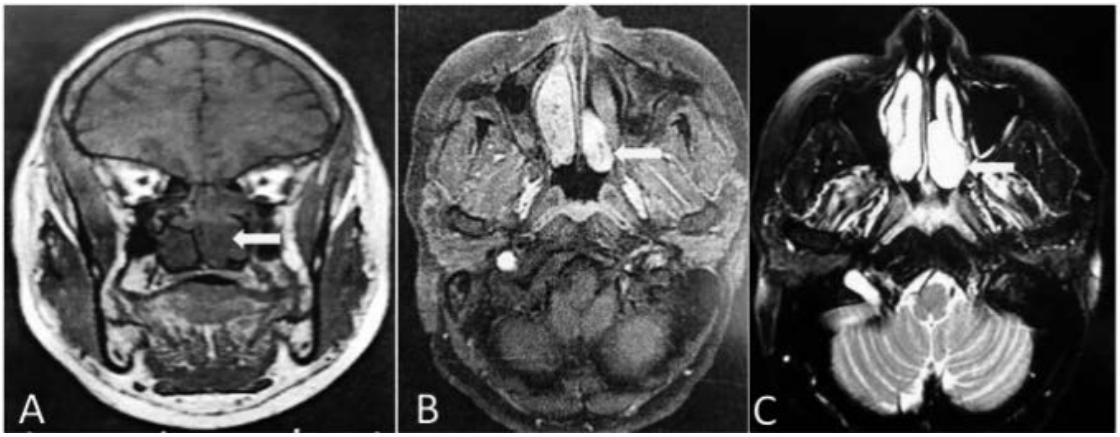

Figure 2 : (A) coronal T1-weighted none contrast MRI, (B) axial T1-weighted post-contrast MRI, (C) axial T2-weighted MRI: a vascular lesion occupying the left nasal cavity mass isointense on T1-weighted, hyperintense on T2-weighted with intense contrast enhancement.

Pre-operative tumor embolization was not performed in light of the small size of the tumor. Considering the size, limited expansion and the location of the tumor, an endoscopic resection was performed. The bleeding was controlled without difficulty by compression with pledgets soaked in diluted adrenalin. Extemporaneous histology examination confirmed the absence of signs of malignancy.

Histological examination showed normal respiratory epithelium with a proliferation of short spindle-shaped cells with slightly branching vascular structures. Stromal bleeding was also noted; however, neither necrosis nor cytologic atypia were observed. The tumor cells were strongly positive for β-catenin and α-smooth muscle actin, and negative to CD34 and Pan-Cytokeratine (Figure 3). These findings were compatible with glomangiopericytoma. During the 12-months follow-up no recurrence was observed.

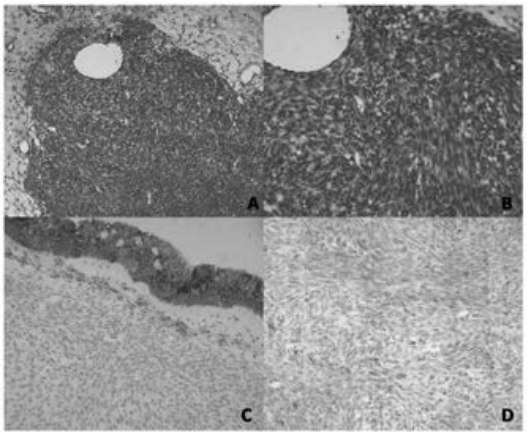

Figure 3 : A (HEx100), B (HE x 200), C (Cytokeratine x 200), D (β-catenin x 200): (A) and (B) tumor was composed of uniform spindle-cells with perivascular arrangement, (C) Negativity of tumoral cells with Pan-Cytokeratine, (D) diffuses expression of tumoral cells with β-catenin.

DISCUSSION

Glomangiopericytoma of the nasal cavity is a rare mesenchymal tumor. it develops in the nasal cavity and/or paranasal sinuses, where it represents less than 0.5% of all sinonasal neoplasm [1]. GPC belongs to the category of borderline and low-malignant-potential soft tissue tumors of the nose and paranasal sinuses.

It can occur at any age; however, previous case series have suggested a peak incidence during the sixth and seventh decades in both genders, though, as in our case, it can occur earlier [1,3,4]. Some series shows a slight female predominance[1,4].

GPC are of unknown etiology. High vascularization caused by previous trauma, hypertension, and long-term steroid use are reported to be possible causes [1].

Clinically, as in our case, patients most often present with epistaxis and/or nasal obstruction.It may, rarely, provoke vision impairment, headache and local swelling because of local infiltration[1,3,5].

On examination, it usually appears as a quite vascular mass that can vary in color, size and consistency. According to some authors, this tumor is predominantly located in the nasal cavity, although some cases in the paranasal sinuses have been reported [6].

The radiological imaging techniques (CT and MRI) can provide adequate information with regards to extension of the tumor. The CT scan shows a soft-tissue mass with strong enhancement after contrast medium. It can demonstrate bone destruction in the sinonasal cavity and adjacent structures [1,7]. On MRI the mass appears solid isointense on T1-weighted images with strong contrast enhancement, while on T2-weighted images the signal is variable [7]. The angiography is used to plan a preoperative embolization [7,8].

According to the literature, the management of this tumor has been achieved through a variety of approaches. The open surgery was the only strategy to achieve free margins [9]. Actually, the endoscopic approach is the treatment of choice with comparable outcomes [1,5,10].

The preoperative embolization prior to the endoscopic excision is recommended in case of a large or highly vascularized tumor [8].
Other treatment modalities such as adjuvant radiotherapy or chemotherapy are still controversial [4,11]. However, in patients with a positive margin, adjuvant therapy could be considered [1,4].
The histological examination is important for the diagnosis. The diagnosis of GPC is often based on the architectural features, especially the capillary pattern. The Hematoxylin–eosin staining shows elongated or ovoid cells, surrounding the normal respiratory epithelium. These tumor cells are characterized by round, punched-out central nuclei and pale eosinophilic cytoplasm [4,7,12].
The immunohistochemical profile of glomangiopericytoma is characterized by diffuse reactivity to vimentin, actin,β-catenin and negative staining for CD34, CD31 and FVIII-Rag [4,10]. In our case, cells were strongly positive for β-catenin and α-smooth muscle actin, and negative to CD34. In addition to its histological and immunohistochemical features, mutations in exon 3 of the gene coding for β-catenin (CTNNB1) and its nuclear expression were recently discovered in GPC [12]. The prognostic factors for aggressive behavior are a large tumor size (> 5 cm), bone invasion, profound nuclear pleomorphism, increased mitotic activity, necrosis, and a higher proliferative index [2].
Life-long follow-up is generally required because of the risk of local recurrence [1]. Complete surgical resection could enhance disease-free survival, and adjuvant treatment could be helpful to prolong survival when complete resection is impossible[1,4].

CONCLUSION

Glomangiopericytoma is rare vascular tumor which diagnosis algorithm involves endoscopy, imaging (CT and MRI) for lesion characterization and extension.
Successful management depends on complete resection. Recent advances in nasal endoscopic surgery have enabled the complete resection of these tumors, minimizing morbidity and facilitating subsequent surveillance of the operative site.

Conflicts of interest: Authors declared no conflicts of interest.

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