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Case report Free Access

Glomangiopericytoma, a rare sinonasal hemangiopericytoma with particular characteristics

Ehsan Arjmandzadeh [1], Abolfazl Taheri [2]*

[1] Shahid Sadoughi University of Medical Sciences, Yazd, Iran

[2] Baqiyatallah University of Medical Sciences, Tehran, Iran

Abstract

Introduction: Glomangiopericytoma (GPC) is an extremely rare paranasal sinuses and nasal cavity vascular neoplasm introduced and differentiated from the conventional hemangiopericytoma in 1998 by Granter et al. Up to now, to the best of our knowledge less than 250 confirmed cases have been reported in the literature. However, the exact etiology is unknown but some risk factors including trauma, hypertension, long term steroid use and pregnancy have been suggested as predisposing factors. Nasal obstruction is the most common presentation followed by intermittent epistaxis, pain, proptosis and epiphora.

Case presentation: Here we describe a case of right sided glomangiopericytoma that was completely resected with safe margins by a Weber-Ferguson approach and underwent adjuvant radiotherapy with no evidence of recurrence one year after surgery.

Discussion: Although glomangiopericytoma is very rare but it should be considered in case of confronting a unilateral vascular mass especially in pregnant females with a history of hypertension, trauma or long-term steroid usage. Definite diagnosis is based on immunohistochemistry and preoperative imaging is mandatory as endoscopic approach should be kept for small sized tumors with definitely identified origin.

Conclusion: Glomangiopericytoma is a rare tumor classified as a low-grade borderline malignancy tumor. Complete excision and long term follow up due to high rate of recurrence are required.

Keywords: Hemangiopericytoma, Paranasal sinuses and nasal cavity, Vascular Neoplasm, Complete Excision

Corresponding Authors: Abolfazl Taheri

✉ Email: dr.abolfazl.taheri@gmail.com

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Introduction

Glomangiopericytoma (GPC), is an extremely rare sinonasal vascular neoplasm which comprises less than 0.5 percent of all sinonasal neoplasia that introduced and differentiated from hemangiopericytoma in 1998 by Granter et al and since then to the best of our knowledge less than 250 confirmed cases have been reported in the literature (1-3). This neoplasm differs from the conventional hemangiopericytoma in three aspects, first the anatomical origin, GPC is usually seen in sinonasal tract, second in biological behavior, GPC as in many reports is an indolent low malignant potential neoplasm with excellent prognosis in case of complete surgical resection but it's potential for local invasion and metastatic spread has been reported too, and finally the third difference is histopathological features considering the fact that GPC originates from perivascular modified smooth muscle cells with round, spindled and focally disposed whirling pattern cells frequently expressing smooth muscle actin (SMA) and CD 34 in immunohistochemistry assessments (4, 5). The tumor is very slightly female dominant and although seen in all age groups, the peak incidence occurs during the fifth and sixth decades of life and is usually presented with unilateral nasal obstruction and/or epistaxis and facial pain or headache (3, 6, 7). In 2005 the WHO classification of head and neck tumors proposed that sinonasal hemangiopericytoma should be named glomangiopericytoma considering their similarity with glomus tumors (8).

Here we describe a case of right-sided glomangiopericytoma, treated surgically with open approach by a typical incision of Weber Ferguson.

Case presentation

A 48-year-old man presented to our medical center with chief complaint of complete right sided nasal obstruction. The obstruction process was progressive with almost six weeks of partial obstruction that converted to complete obstruction from two months ago. He felt a radicular sharp right hemifacial pain about two weeks before the onset of right nasal obstruction causing extraction of first premolar and molar teeth of right upper jaw due to severe pain without prominent pain relief. Concomitant with complete right nasal obstruction he noticed non-tender stiff bulging of right hard palate. Also, the patient complained of ipsilateral anosmia epiphora and intermittent epistaxis.

Patient's past medical history showed chronic rhinosinusitis and he had a surgical history of septoplasty about twelve years ago. There was no history of addiction to tobacco or other drugs.

On examination right sided facial tenderness was detected and oral examination showed a non-tender compressible mass located on the right side of hard palate which was exceeded from the midline (Figure 1).

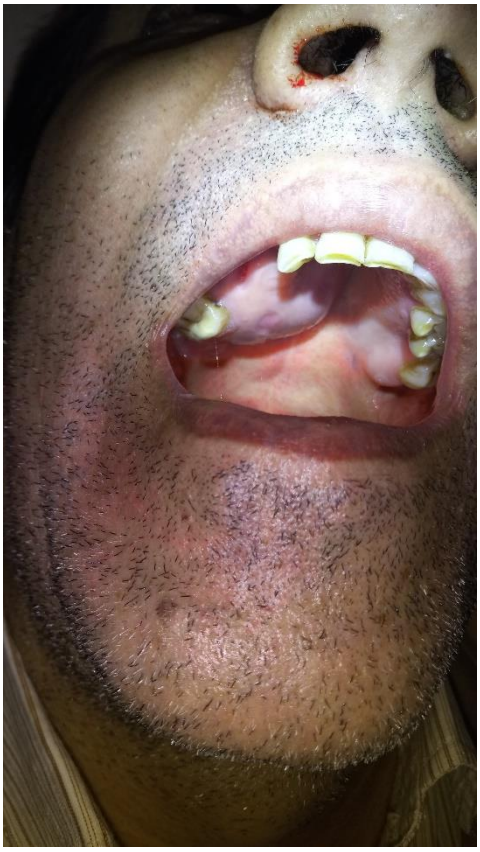

Figure 1. Extension of the mass into oral cavity presented as right hard palate bulging.

Anterior rhinoscopy and rigid nasopharyngoscopy revealed a fleshy, greyish pink polypoid mass that bled easily with minimal manipulation.

No enlarged cervical lymph nodes were palpated.

Computed tomography (CT) scan showed a right maxillary sinus soft-tissue density mass which was destructive in nature with maximal length of about seven centimeters occupying the whole right maxillary sinus and ipsilateral nasal cavity extending to oral

cavity with bony destruction of medial maxillary sinus wall, right nasal turbinates, nasal septum and right hard palate but sparing orbital floor.

Magnetic resonance imaging (MRI) with and without gadolinium demonstrated a T_1 hypointensity and T_2 hyperintensity with a bright mass on T_1 with contrast within right maxillary sinus and nasal cavity extending to ethmoid cells superiorly and oral cavity inferiorly (Figure 4b).

Histopathological examination with immunohistochemistry (IHC) study showed bundles of spindle cells proliferation with atypia and positive for SMA, CD_{31}, BCL_2 and EMA with hemangiopericytoma- like pattern suggestive of glomangiopericytoma (Figure 2).

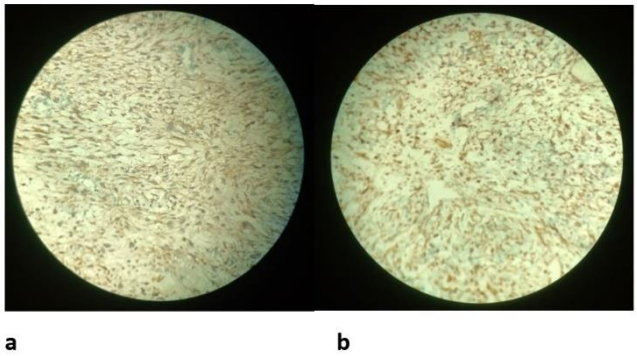

Figure 2. Muscle actin (SMA) as seen above (a) and B cell lymphoma 2 (BCL_2) markers (b).

The patient underwent a right open subtotal maxillectomy approach with Weber-Ferguson incision and the tumor was resected completely (Figure 3) with safe margins and for better local control, bony boundaries invaded by the tumor were drilled too.

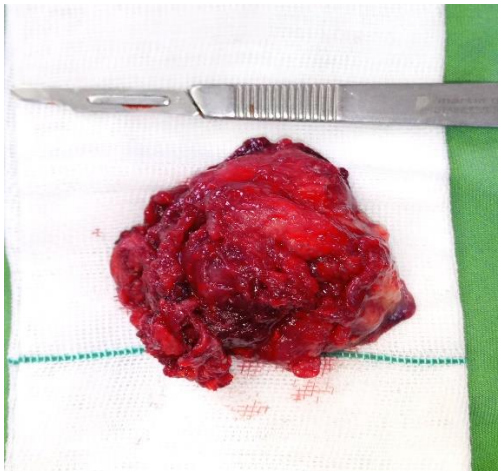

Figure 3. A spongy compressible mass was resected completely from right maxilla and nasal cavity with maximal diameter of 7 cm.

Finally, a full-thickness skin graft was harvested from anterolateral part of right thigh and transplanted to the posterosuperior and anterior walls of the space helping better mucosalization of the cavity.

Also, a hard palate obturator prosthesis was used to close the palatal defect.

The operation was followed by intensity-modulated radiation therapy (IMRT) over right maxilla in 30 fractions and for a total of 65 Gy. Our patient remained free of local or regional recurrence after one year post operation as followed by regular visits and CT scans (Figure 4a).

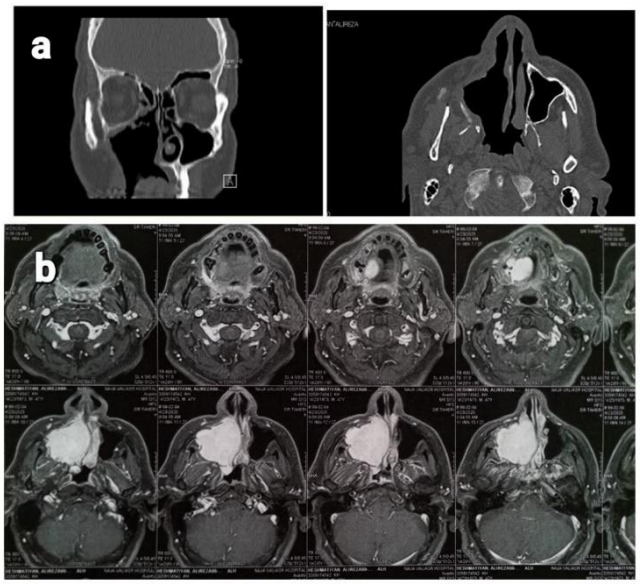

Figure 4. Coronal and axial view of paranasal sinuses CT scan one year post operation, No evidence of recurrence is seen (a). Axial T_1 with gadolinium injection before surgery demonstrates a bright mass (b).

Discussion

Glomangiopericytoma is a kind of hemangiopericytoma found in head and neck especially sinonasal tract originating from pericytes attached on the surface of capillaries acting as sphincters with the function of blood flow control (9).

Hemangiopericytoma is considered a rare soft tissue tumor usually (85%) found in retroperitoneum and lower extremities and less common (15%) in head and

neck region which is known as glomangiopericytoma (9, 10). Most common sites of involvement in head and neck are scalp, face, neck, nasal cavities and paranasal sinuses (11). Nasal cavities are twice more involved and between paranasal sinuses, involvement of ethmoid cells and sphenoid sinuses with glomangiopericytoma is about four times more often seen than it does in maxillary sinuses (12). In our case we could not determine the exact origin of tumor as the whole right sinonasal cavity was involved by the tumor.

Gender distribution is almost equal although some studies report a slight female dominancy (6) contrary to our case who was a man. It is commonly seen in the sixth and seventh decades of life and more than 80% of patients are Caucasian (3, 7) but our patient was just 48 years old and the tumor appeared at least one decade earlier. However, the exact etiology is unknown but some risk factors including trauma, hypertension, long term steroid use and pregnancy have been suggested as predisposing factors (3, 6, 7, 12). The patient we reported here had none of the risk factors.

Glomangiopericytoma is considered as a painless very slow growing mass and hence found in large sizes when medical diagnosis is made (13). Nasal obstruction is the most common presentation (60%) followed by intermittent epistaxis (50%), pain, proptosis and epiphora (11). Our patient's chief complaint was unilateral nasal obstruction and facial pain. On rigid endoscopy glomangiopericytoma is appeared as a soft, fleshy, and red to greyish pink mass which is edematous to hemorrhagic and easily bleeds (14). There is no relationship between tumor behavior and anatomical site in the literature but generally about 10% of the cases will finally encounter distant metastasis through hematogenous route and usually to lungs, bones and liver and up to 40% will have local recurrence (4, 11).

As biopsy is not recommended due to severe bleeding, complete radiological examination by CT and MRI should be performed. A CT scan shows a soft tissue mass which possible bone destruction is clearly demonstrated and in case of contrast administration strong enhancement is seen (6, 15). Because of poor ability of CT scan to differentiate between mass and inflammatory fluid MRI is mandatory as well (6). On T_1 weighted MRI, glomangiopericytoma appears as a solid hypointense to isointense mass with bright enhancement after intravenous contrast injection and on T_2 weighted imaging, contrary to inflammatory fluid, glomangiopericytoma appears as a moderate to low intensity mass (14). The patient had preoperative CT imaging which consistently demonstrated non-specific findings of mass-like lesion of the para-nasal sinuses extending up toward the skull base. MRI with contrast administration and in-office endoscopy showed a vascular and easily bled mass.

Estimating the risk of clinical aggressiveness is very hard and although a certain potential of malignancy should always be taken into account, several malignancy criteria have been established including histological necrosis, nuclear atypia, high number of mitosis and a large tumor size of >6.5 cm (5, 9).

Diagnosis of glomangiopericytoma is based on histopathology. Hematoxylin and eosin staining reveals many vascular vessels and perivascular hyalinization with uniform oval shaped cells and round to spindle shaped nuclei but it is immunohistochemistry studies that can differentiate soft tissue hemangiopericytoma from sinonasal hemangiopericytoma by characteristic reaction for actin and vimentin (1, 2, 4). Immunohistochemistry evaluation in our patient was consistent with positive muscle actin (SMA) and B cell lymphoma 2 (BCL_2) markers which are charecteristic for GPC.

The treatment of choice is wide local excision which is done in two main endoscopic and open approaches. Considering the fact that completeness of resection is known as the main predictor of recurrence, the literature advices endoscopic resection to be kept for those tumors with small size, definitely identified site of origin and tumors undergone preoperative embolization with Onyx although patient's preference and surgeon's technical expertise should be considered when final decision is to be taken (6, 9). Due to large size of our patient's mass and inability to determine the origin of the mass by endoscopic approach he went under an open surgical approach. Radiotherapy as a primary treatment modality has a recurrence rate as high as 50% but when used as an adjuvant therapy in case of incomplete resection of the tumor it had a statistically lower rate of recurrence but no difference

in overall survival rate (1, 6, 9, 11, 13). Although there are some chemotherapy agents used as trial studies, currently there is no evidence for or against the use of chemotherapy agents in hemangiopericytoma or hemangiopericytoma-like tumors including glomangiopericytoma (1, 6, 11, 16-18). Our patient received appropriate adjuvant radiotherapy and he was free of tumor residue or recurrence up to one year of follow-up.

Conclusion

Sinonasal hemangiopericytoma or glomangiopericytoma is considered as a rare sinonasal tumor and differs from usual somatic hemangiopericytoma in the anatomical site of tumor origin, biological behavior and histopathological properties. It is classified as a low-grade borderline malignancy tumor and should be considered when imaging and endoscopic evaluations show a soft tissue polypoid vascular nasal cavity mass. Complete excision of the tumor is the treatment of choice. Due to the high rate of recurrence even 17 years after the initial presentation, long-term follow-up is required (Table 1).

Table 1. Glomangiopericytoma (summarized).

Clinical features	Risk factors	Imaging findings	IHC findings	Treatment
Unilateral nasal obstruction	Pregnancy	Soft tissue mass in CT scan	SMA +	Wide complete resection ± radiotherapy
Epistaxis	Trauma	Bright signal in MRI	BCL2+	
Headache or facial pain	Long term steroid			

Future research

Future studies are needed to clarify the most important risk factors and the exact pathogenesis and also to standardize the criteria for adopting the best surgical approach (endoscopic or open) and post-operative follow-up.

Author contribution

EA and **AT** contributed to data gathering involved in drafting the manuscript, **EA** drafted the initial manuscript and **AT** provided a review of the manuscript. Both authors approve of the final manuscript.

Conflict of interest

The authors declare that they have no conflict of interest.

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