CASE REPORT

Glomangiopericytoma: A rare tumour of sinonasal cavity

Shayan Khalid Ghaloo[1], Rahim Dhanani[2], Hamdan Ahmed Pasha[3], Muhammad Wasif[4], Saira Fatima[5], Mubasher Ikram[6]

Abstract

The history of glomangiopericytoma began in 1924 when it was initially diagnosed by Stout and Murray. It is a rare tumour of the respiratory mucosa, with a prevalence of less than 0.5% among all sinonasal tumours. Literature shows female predominance among patients who develop glomangiopericytoma. So far, no accurate aetiology has been discovered, but there are certain risk factors, including trauma, use of corticosteroids and high blood pressure, which are believed to cause glomangiopericytoma. Patients usually present with a history of epistaxis or nasal blockage, though symptoms can get worse if the tumour is not resected timely and can lead to visual disturbance, chronic sinusitis and headache. It has reddish polypoidal appearance on examination. The best modality for the treatment of glomangiopericytoma is endoscopic surgical resection via trans-nasal approach. We present the case of a 70-year-old man, with nasal blockage and epistaxis who underwent endoscopic sinus surgery for this condition.

Keywords: Glomangiopericytoma, Nasal tumor, Epistaxis, Nasal obstruction. Endoscopic sinus surgery.

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Introduction

Glomangiopericytoma (GPC), also known as sinonasal-type haemangiopericytoma, is a rare mesenchymal vascular tumour that arises from the pericytes surrounding the capillaries.[1] It was categorised as a borderline and low-malignant potentially soft-tissue tumour of the nose and paranasal sinuses by the World Health Organisation in 2005, and accounts for less than 0.5% of all sinonasal tumours.[2] Glomangiopericytoma was initially diagnosed by Stout and Murray in 1942 and was defined as haemangiopericytoma.[3] Since its initial description, the definition of the disease is surrounded by controversy and has been questioned.[4] In 1976, it was described by Compagno as 'haemangiopericytoma like' due to the low incidence of metastasis and mortality.[3] The aetiology of this disease is still not clear though trauma, hypertension and use of steroids are considered the predisposing factors that may cause its occurance.[5] We present a rare case of GPC in a 70-year-old male presenting to us in the clinic with unilateral nasal obstruction and epistaxis, and was subjected to endoscopic sinus surgery.

[1-4,6]Department of Surgery, Aga Khan University Hospital, Karachi, Pakistan; [5]Department of Pathology, Aga Khan University Hospital, Karachi, Pakistan.
Correspondence: Rahim Dhanani. e-mail: dr.rahimdhanani@gmail.com

Case Report

A 70-year-old male with no known comorbidity presented in the outpatient department of Aga Khan University Hospital, Karachi, a tertiary healthcare centre in February 2019 with a history of progressive right nasal obstruction for one year and epistaxis for one month. On anterior rhinoscopy, there was staining of nasal mucosa with blood which correlated with the history of epistaxis. On examination, nasal patency showed a patent left and obstructed right nasal cavity.

Endoscopic nasal examination revealed a reddish, polypoid and vascular mass in the right nasal cavity. It was located over the middle turbinate, adherent to the nasal septum, while the posterior extent could not be evaluated. Nasal mucosa was intact and no ulceration or erosion was noticed.

Computed tomography (CT) scan revealed an enhancing lesion in the right nasal cavity posteriorly, measuring 16 x 10 mm. The lesion lay within the anterior ethmoid air cells, adherent medially to the nasal septum and laterally to the right lateral wall of the nasal cavity. (Figure 1). It was seen obstructing the passage of right frontal and ethmoid sinuses. Magnetic resonance imaging (MRI) was not done as there was no intracranial extension.

Biopsy revealed features consistent with GPC and endoscopic removal under general anaesthesia was planned. The patient underwent removal of the right nasal mass through endoscopic sinus surgery. Intraoperatively, it was a 1.5 x 1 cm polypoid lesion lying posteriorly in the nasal cavity and closely adherent to the middle turbinate laterally and nasal septum medially. The blood loss, measured by counting output in the suction pump, was less than 100 ml, and the operation time was 35 minutes. It was a day care surgery, and the patient was discharged on the same day after removal of nasal packing that was kept in place for three hours.

Histopathological specimen contained multiple polypoid and smooth-surfaced tan brown hyperaemic irregular tissue fragments measuring 3 x 2 cm in aggregate.

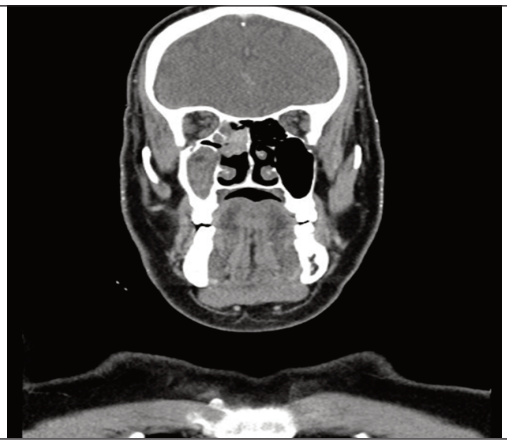

Figure-1: CT scan coronal view showing an enhancing lesion in the right nasal cavity posteriorly, measuring 16 x 10 mm.

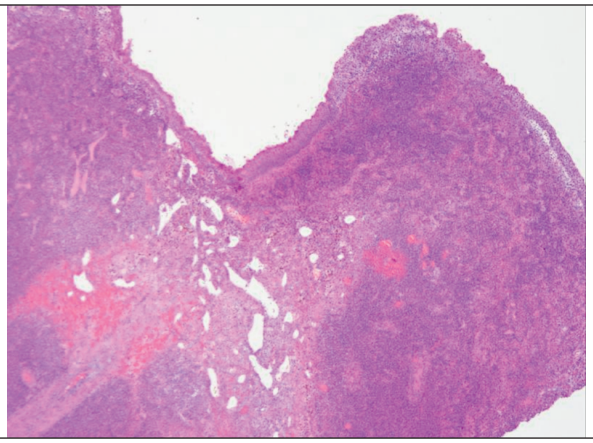

Figure-2: Nasal mucosa with underlying spindle to polygonal cells with prominent vasculature.

Histological examination revealed nasal mucosa with underlying spindle to polygonal cells with prominent vasculature. (Figure 2). Pericytic cells were seen emanating from the vessels. Bland round nuclei of the cells were also noted on the microscopic examination. Immunohistochemical stains were positive for ASMA (Alpha Smooth Muscle Actin) and negative for CD-34. (Figure 3, 4). The histopathological features were compatible with GPC of the sino -nasal cavity.

On three-months follow up visit, the patient was symptom-free with no active complaints. Endoscopic examination showed no signs of recurrence. No complications were noted post-operatively.

Discussion

GPC is an extremely rare mesenchymal tumour almost exclusively arising from the sino-nasal cavity and is characterised by prominent perivascular growth.[1] It was diagnosed by Stout and Murray who defined it initially as haemangiopericytoma but later, in 1976, Compagno

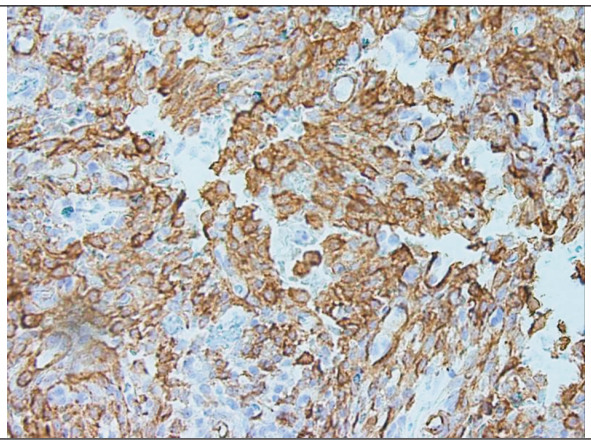

Figure-3: Immunohistochemistry showing positive ASMA staining.

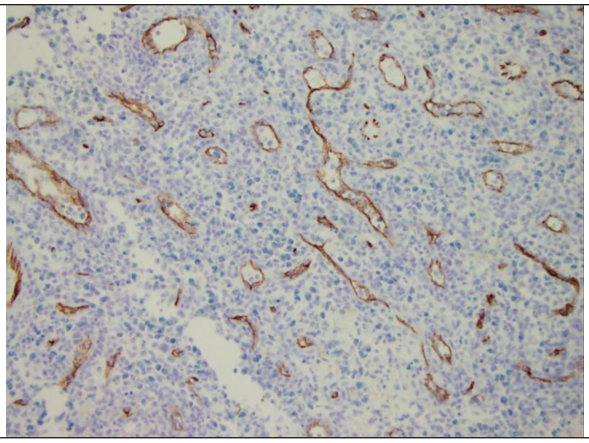

Figure-4: Immunohistochemistry showing negative CD-34 staining.

defined this entity as 'haemangiopericytoma like' due to the low incidence of metastasis and mortality.[3]

It comprises less than 0.5% of all sino-nasal neoplasia.[2] The median age of the patients is 60 years and ranges from as low as second decade of life to 86 years. Most cases reported in literature suggest that it is a female predominant disease.[6] Contrastingly, here we report GPC in a 70-year-old male.

Patients most commonly present with epistaxis (78%), followed by other symptoms, that may or may not be there, such as nasal obstruction (52%) and headache (17%).[7] In our case, the presenting complaint was prolonged nasal blockage and epistaxis.

These tumours arise from the respiratory mucosa of nasal cavity. Some tumours, however, if not managed early, can extend and involve one or more of the sinuses and, in advanced stage, it can also spread to the skull base region. In those advanced stage tumours, the symptomatology may be different, and the patient may present with visual disorders, chronic sinus condition or paraesthesia of the

cheek.[8]

Clinically, the tumour may appear to be submucosal, beefy red, soft and haemorrhagic without any mucosal ulceration. CT invariably shows a soft-tissue mass with strong enhancement in the unilateral nasal cavity or paranasal sinuses.[2]

The aetiology of this tumour is still not clear. The risk factors which may have a role in the aetiology include previous trauma, high blood pressure, prolonged use of corticosteroids, pregnancy, and hormonal imbalance.[9] No such risk factors were noticed in our patient. Differential diagnosis of GPC may include Glomus tumours, Desmoid-type fibromatosis, and nasopharyngeal angiofibroma.[2] GPC is diagnosed by histological characteristics showing epithelioid cells in a perivascular pattern with frequent perivascular hyalinisation. Tumour cells are immune-histologically positive for cytoplasmic SMA and Vimentin, and nuclear β-catenin in 80–100% cases. Tumour cells exhibit no strong diffuse staining for CD34 and are basically negative for AE1/AE3, Bcl-2, CD34, CD99, CD117, Factor VIIIR Ag, S-100protein, and STAT6.[10]

Complete surgical resection is the standard treatment in GPC because the tumour is relatively resistant to chemotherapy and radiation.[11] As the tumour is highly vascular, some authors have also advocated role of preoperative angioembolisation.[12] We didn't opt for preoperative angioembolisation as the tumour was small in size and the epistaxis was not severe. We were able to completely excise the tumour through endoscopic sinus surgery.

Although WHO has characterised it as a low malignancy tumour, in a few cases it can recur and prove to be fatal.[1] Recurrence can be a consequence of incomplete excision. In our case, the tumour was completely excised and, hence, the potential of recurrence was low; however, long-term follow up is required. We donot have any clinical picture of this mass, which is the limitation of this paper.

Conclusion

GPC is a rare tumour that arises from the respiratory mucosa. We reported case of a 70-year old male who presented with progressive unilateral nasal obstruction and epistaxis. Biopsy was performed to determine the diagnosis. CT scan was done to see the complete extent of the tumour. Complete surgical excision was undertaken, which is currently the treatment of choice for GPC.

Disclaimer: Patient's consent was taken to report this case.

Conflict of interest: None.

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