Sinonasal Glomangiopericytoma

CASE REPORTS

Sinonasal Glomangiopericytoma - A Case Report

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ABSTRACT

Sinonasal glomangiopericytoma is a tumor showing similarities to a glomus tumor but is less aggressive with a low malignant potential. The treatment of choice is complete excision followed by a regular long-term surveillance. The role of adjuvant chemotherapy and radiation remains undetermined. Our patient was a 45-year-old man who presented with nasal obstruction. Endoscopic excision showed sinonasal glomangiopericytoma. No adjuvant treatment was offered and patient was advised active surveillance. Patient was on a regular three-month follow-up. There was no evidence of disease recurrence after one year.

Keywords: Glomangiopericytoma, Hemangiopericytoma, Sinonasal neoplasms.


INTRODUCTION

Glomangiopericytoma is a sinonasal tumor, having a perivascular myoid phenotype, characteristic vascular proliferation and small vessel perivascular hyalinization. It was initially described in 1942, by Stout and Murray. The World Health Organization (WHO) has classified this tumor as glomangiopericytoma since 2005. It is a rare disease which accounts for <0.5% of all the sinonasal tumors. The etiology is not known although history of previous trauma, pregnancy, hypertension, corticosteroid use and occupational exposures might lead to the increased incidence. The role of radiotherapy and chemotherapy are still undetermined.

CASE REPORT

Our patient, 45 years of age, resident of Kotli Azad Kashmir Pakistan, presented to Combined Military Hospital (CMH) Rawalpindi, with one-month history of nasal discharge, nasal obstruction especially at night and feeling of foreign body in the nose. He had frequent nasal discharge and off and on blood stained sputum. He also had one episode of epistaxis after manual injury. He visited Otorhinolaryngology Outpatient Department at CMH Rawalpindi. His examination revealed a polypoid mass in the left nares. CT scan face and neck showed a well-defined, enhancing mass lesion of 2.7 x 1.7 x 1.9 cm (AP <Tr, CC), in left half of nasal cavity, encroaching osteo-meatal complex and invading middle and inferior turbinates and erosion of bony part of nasal septum (Figure).

The patient was counselled for different excision options. He chose the endoscopic resection. Examination under anesthesia revealed a bleeding mass arising from mid-point of inferior turbinate. Excision of the mass was carried out by Fiber-optic Endoscopic Sinus Surgery (FESS) along with type II left maxillary antrostomy. Fungus filled maxillary sinus was washed and hemostasis was secured. A polypoid mass (3x2.4x1cm) was excised. Post-operative recovery was uneventful. Histopathology came out to be sinonasal glomangiopericytoma. On immune-histochemistry, SMA, beta-catenin and cyclin D1 were stained positive. Markers of STAT 6 and CD-34 were negative. The patient then visited Clinical Oncology Out-Patient Department. On examination, he had no residual or recurrent disease. His contrast enhanced CT scan of face and neck showed no abnormality. This case was discussed at head and neck oncology tumor board. No adjuvant treatment was offered. Patient was put on follow-up. After
one year of regular three monthly follow-up, patient is currently disease free.

**DISCUSSION**

Hemangiopericytomas originate from pericytes. These occur particularly in the nasal cavity and paranasal sinuses. Glomangiopericytoma has a low probability of developing into malignancy. It differs from conventional soft tissue hemangiopericytoma in location, histology and biologic behavior, with lower rates of metastasis and mortality. There is presence of strong nuclear β-catenin expression which supports a mutation in the CTNNB1 gene.

Clinically it resembles the allergic polyp and is mostly unilateral and unencapsulated with an average size of around 3cm. It may provoke pain because of local infiltration, headache, visual impairment and local swelling. The hemorrhagic sinonasal mass has multiple differential diagnoses including solitary glomus tumors, glomangioma, conventional hemangiopericytoma, myopericytoma and angioleiomyoma. Glomangiopericytomas with aggressive behavior are rare and usually have larger size (>5 cm), profound nuclear pleomorphism, invasion of bone, necrosis and increased mitotic activity (>4 per 10 h.p.f).

Endoscopic surgery is favored, giving a better-magnified sight, with minimal blood loss. Incomplete excision may increase the chances of recurrence. Radiotherapy and chemotherapy are used in cases of inoperable tumors, metastasis or as a palliative therapy. Radiotherapy following an incomplete surgical resection decreases the rate of recurrence. Post-radiotherapy rhinosinusitis is treated with intra-nasal inhalational steroids. Salvage surgery can be done in case of recurrence.

In a nutshell, definitive surgery is sufficient for the treatment of sino-nasal glomangiopericytomas. Radiotherapy and chemotherapy are reserved for unresectable disease. Close follow-up and surveillance of patient after surgery should be done, for the early detection of recurrence and provision of appropriate salvage treatment as recommended by multi-disciplinary tumor boards.

**Conflict of Interest:** None.

**Authors’ Contribution**

MSN: Principal investigator, conceptualisation and manuscript writing, UA: Concept & data collection, AZ: Data analysis, MYK: Interpretation of data.

**REFERENCES**