DOI: 10.14260/jemds/2014/3995

CASE REPORT

NASAL MASS: AN ENIGMA

Somu L[1], Prasanna Kumar S[2], Ravikumar A[3], Raadhika Shree[4]

HOW TO CITE THIS ARTICLE:
Somu L, Prasanna Kumar S, Ravikumar A, Raadhika Shree. "Nasal Mass: An Enigma". Journal of Evolution of Medical and Dental Sciences 2014; Vol. 3, Issue 69, December 11; Page: 14850-14856,
DOI: 10.14260/jemds/2014/3995

ABSTRACT: Glomangiopericytoma is an uncommon sinonasal neoplasm, originating from pericytes of capillaries constituting less than 1% of all sinonasal mass. It is a borderline low malignant tumor. We report the case of a 25 year old female with history of progressive nasal obstruction on the left side, bleeding from the left nasal cavity, anterior rhinoscopy showed a mass in the left middle meatus. This was successfully excised in toto by trans-nasal endoscopic approach. Histopathology was inconclusive and Immuno-histochemistry was required to confirm the diagnosis. The clinical presentation, histopathology, management and prognosis are discussed.

KEYWORDS: Glomangiopericytoma, nasal tumor, endoscopic transnasal surgery.

INTRODUCTION: Glomangiopericytoma (Sinonasal-type hemangio-pericytoma) is an uncommon, borderline low malignant potential soft tissue tumour, constituting less than 1% of the tumours of the nose and paranasal sinuses.[1,2,3] It is defined by WHO as a sinonasal tumor that demonstrates a perivascular myoid phenotype.[4] It was first described by Stout and Murray in 1942.[5] It differs from hemangiopericytoma in location of occurrence, biologic and histologic properties.[6]

Complete surgical excision is the treatment of choice. Endoscopic transnasal approach gives an elegant and precise surgical excision of the tumor of the nose and paranasal sinus without any cosmetic defect.

CASE REPORT: A 25 year old female presented to our OPD with history of progressive left nasal obstruction and recurrent unprovoked bleeding from the left nasal cavity for 3 months. She also complained of recurrent headache over the left frontal region associated with upper respiratory tract infections.

Anterior rhinoscopy showed a reddish mass occupying the region of left middle meatus and middle turbinate. The mass was not sensitive to touch and was bleeding on touch or probing. The left frontal sinus tenderness was present. The skin of the external nose was normal and there were no palpable cervical lymph nodes. Diagnostic nasal endoscopy showed a pink globular mass occupying the entire middle meatus region extending to base of skull superiorly, impinging on the septum medially. (Figure 1).

DOI: 10.14260/jemds/2014/3995

Figure 1: DNE picture showing a reddish mass occupying the region of left middle meatus and middle turbinate.

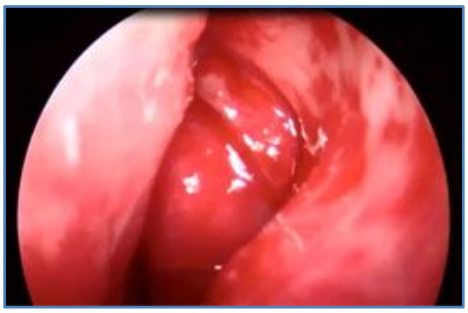


Fig. 1

CT scan with contrast of the paranasal sinus showed a homogenous soft tissue density in the region of the middle meatus extending medially to the septum, superiorly to the skull base, posteriorly to ethmoidal sinuses, laterally breeching the lamina papyraceae of the orbit, contrast enhancement was seen in this lesion (Figure 2).

Figure 2: CT PNS coronal section showing a homogenous soft tissue shadow occupying the middle meatus extending above to the fovea ethmoidalis and laterally into the orbit.

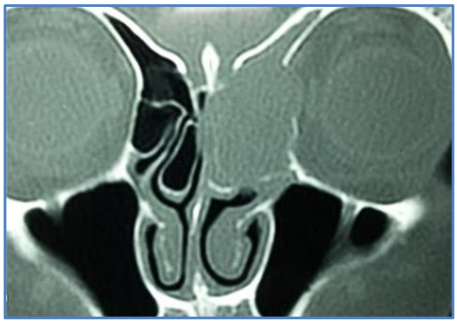


Fig. 2

Figure 3 a: MRI T1 weight coronal section showing low intensity soft tissue signals occupying the region middle meatus extending above to the fovea ethmoidalis and laterally into the orbit.

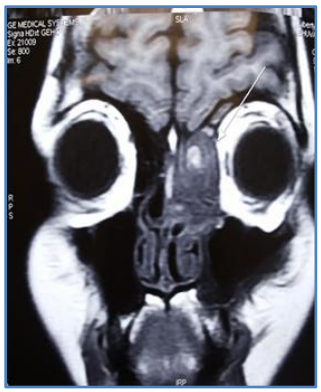


Fig. 3a

DOI: 10.14260/jemds/2014/3995

Figure 3b: T2 weighted MRI Coronal section showing high intensity soft tissue signals occupying the region middle meatus extending above to the fovea ethmoidalis and laterally into the orbit, the lesion is extra dural.

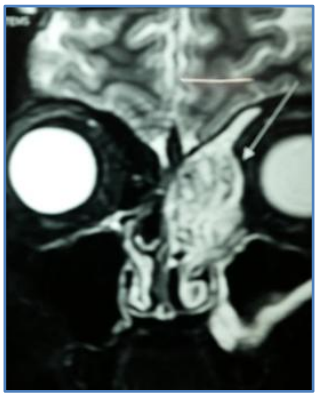


Fig. 3b

Figure 3c: MRI T2 weighted-Sagittal section showing the extension up to the skull base but extradural.

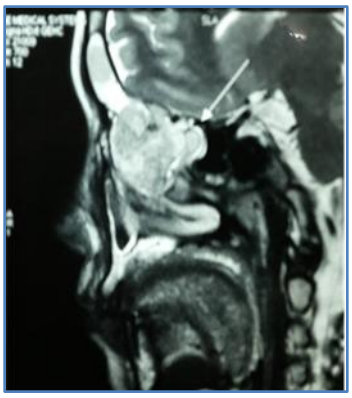


Fig. 3c

Figure 3d: MRI- Axial section showing tumor extension beyond lamina papyracea.

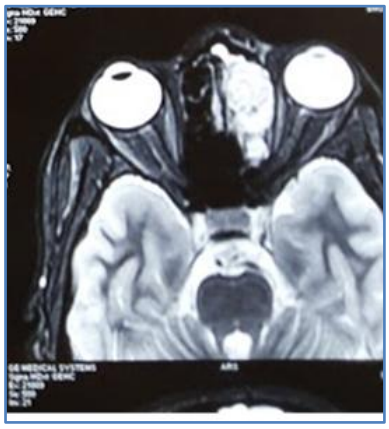


Fig. 3d

DOI: 10.14260/jemds/2014/3995

MRI T1 weighted image (Figure 3a) revealed low intensity lesion and T2 weighted image (Figure 3b) high intensity lesion, occupying the middle meatal region, impinging on the septum medially, extending into ethmoidal sinuses posteriorly (Figure 3c), and breaching the lamina papyraceae of the orbit laterally (Figure 3d).

Biopsy was taken from the mass, and was reported as Glomangiopericytoma. Under General anesthesia the mass was excised in toto via the transnasal endoscopic approach along with the entire lamina papyracea, the periorbita was not involved and hence left alone (Figure 4). Intraoperatively the blood loss was minimal and easily controlled. The excised tissue was sent for HPE. Post operatively this patient was followed up for 1 year with no evidence of recurrence.

Figure 4: Gross image of the lesion excised in Toto.

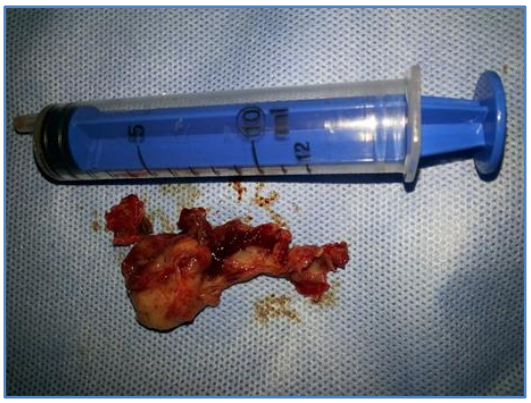


Fig. 4

Grossly it was a reddish mass of 3x5cms with cut surface showing grey white and grey black areas. (Figure. 4)

Histopathological examination showed round to oval cells with oval to spindle shaped nucleus and mild nuclear pleomorphism arranged in sheets surrounding blood vessels. The blood vessels were thin walled. Focal areas of necrosis (10%) and increased mitotic activity (6/10 HPF) was seen. (Figure. 5 a and Figure 5 b)

Immunohistochemistry of the tumor cells were positive for vimentin and Bcl-2 and CD 34 (Figure 5c). They were negative for S 100, Synaptophysin and Chomogranin. Ki-67 labeling index - 20%. (Figure 5d).With the above findings a diagnosis of Glomangiopericytoma was confirmed.

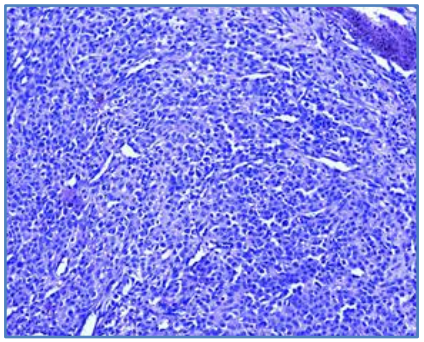
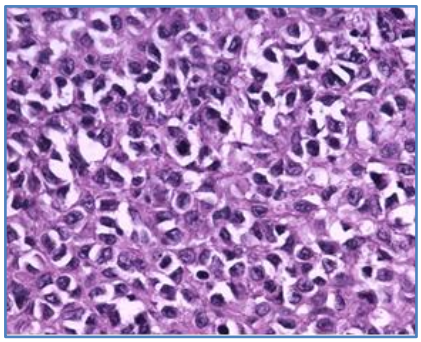
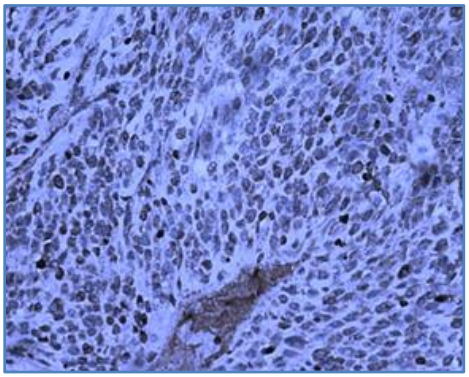


Fig 5a: Histopathology (40 x)

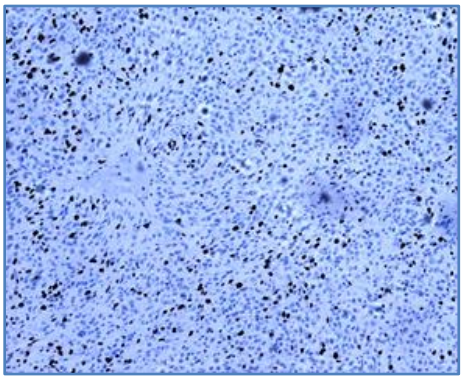


5b: Increased Mitotic activity

DOI: 10.14260/jemds/2014/3995



5c: Bcl2 IHC positive



5d: Ki 67 index >20%

DISCUSSION: Glomangiopericytomas constitute less than 1% of sinonasal tumours, although isolated paranasal sinus involvement has been reported.[1,2,3,4] It was initially thought to fall in the spectrum between glomus tumour and capillary hemangioma.[7] Campagno and Hyams in 1976 classified them as "Hemaniopericytoma-like" tumours.[1,6] The sinonasal variety of hemangioperiytoma behaves in indolent fashion, and shows marked pericyticdifferentiation4and has distinct morphological entities.[7] It differs from hemangiopericytoma in location, biologic behavior and histopathology.

No specific age predilection has been associated with this tumor, this tumor can occur at any age, with incidence being highest in the seventh decade and a with a slight female predominance.[8]

This tumor originates from perivascular glomus like myoid cells although no proof of origin exists. They have a borderline to low malignant potential. It is a soft tissue mass with vascular proliferation including branching vessels and small vessel peri vascular hyalinization with marked morphological heterogenesity.[9,10] In a study by Angouridakis et al, the proposed etiology was due to post trauma, hypertension, pregnancy, and use of steroid.[11]

Clinically the mass looks polypoidal, beefy red to greyish pink, soft, fleshy to friable, edematous to hemorrhagic, and bleeds on touch.[12] Regional lymph nodes are rare.[13]

Histopathology shows diffuse subepithelial well delineated but un encapsulated cellular tumour[14] with proliferation of closely packed spindle cells in variety of pattern which includes diffuse, whorled and storiform, with characteristic hemangio-pericytoma like thin walled vessels with branching staghorn pattern and frequent perivascular hyalinization of capillary sized vessels. In addition to spindle cells, occasionally a focus of round cell appearance of glomus tumour is present. Unlike glomus tumor eosinophilic cytoplasm of Glomangiopericytoma results in synctitial appearance as the cells lack cell borders.[15] The neoplastic cells are uniform, elongated to oval with spindle shaped nucleus and slightly eosinophilic cytoplasm.[12,14,15,17] Mild cytological atypia and mitotic figures may be present.[16] Inflammatory infiltrate with eosinophils, mast cells, lymphocytes and plasma cells may be present. Immunohistochemistry is positive for muscle specific and smooth muscle actin, CD 34, Factor XIII A and vimentin and negative for desmin, cytokeration and S-100, whereas hemangiopericytoma is negative for CD 34 and vimentin.

They are of low malignant potential, but large tumors more than 5 cms, that invade bone, with nuclear pleomorphism, high mitosis (>4/10 hpf) to proliferation index (>10% ki67 index) tend to be aggressive. Glomangiopericytoma rarely warrants aggressive treatment beyond local excision.5 year

survival rate following complete excision is more than 90%. The overall local recurrence following surgical excision is 16.8% as a consequence of incomplete excision and is considered residual disease.[17] Regular post-operative follow up is required.

The differential diagnosis includes sinonasal polyposis, Pyogenic granuloma, Leiomyomas, Angiofibromas, inverted papilloma, Solitary fibrous tumour. This case is reported for its rarity and to emphsise the option of transnasal endoscopic removal of this tumor

CONCLUSION: Glomangiopericytoma is a rare (<1%) sinonasal neoplasm with borderline low malignant potential with little recurrence if excised completely. We have managed the case of a 25 year old female diagnosed clinically and histopathologically as Glomangiopericytoma with complete transnasal endoscopic excision and no recurrence in a period of 1 year.

REFERENCES:

1. Compagno, J. Hemangiopericytoma-like tumors of the nasal cavity: a comparison with hemangiopericytoma of soft tissues. Laryngoscope 1978. 88 (3):460–469.
2. Thompson, L. D., M. Miettinen, and B. M. Wenig. Sinonasal-type hemangiopericytoma: a clinicopathologic and immunophenotypic analysis of 104 cases showing perivascular myoid differentiation. Am J Surg Pathol 2003. 27 (6):737–749.
3. Kuo, F-Y., H-C. Lin, H-L. Eng, and C-C. Huang. Sinonasalhemangiopericytoma-like tumor with true pericyticmyoid differentiation: a clinicopathologic and immunohistochemical study of five cases. Head Neck 2005. 27 (2):124–129.
4. Barnes, L., J. W. Eveson, P. Reichart, and D. Sidransky. eds. Pathology and Genetics of Head and Neck Tumours. Lyon, France IARC Press. 2005. 43–44. World Health Organization Classification of Tumours; Vol 9.
5. Stout, A. P. and M. R. Murray. Hemangiopericytoma: a vascular tumor featuring Zimmerman's pericytes. Ann Surg 1942. 116 (1):26–33.
6. Glomangiopericytoma of Nasal Cavity: A Rare Sinonasal Perivascular Tumor; Chien-Chung Wang, and Sau-Tung Chu; J Med Sci 2013; 33 (2): 107-111. http://jms.ndmctsgh.edu.tw/3302107.pdf DOI: 10.6136/JMS. 2013.33 (2).107
7. Dandekar M, McHugh JB. Sinonasalglomangiopericytoma: case report with emphasis on the differential diagnosis. Arch Pathol Lab Med 2010; 134: 1444-1449. doi: 10.1043/2010-0233-CR.1.
8. Lester D, Thompson M. Sinonasal tract glomangio- pericytoma (hemangiopericytoma). Ear Nose Throat J 2004; 83: 807.
9. Nunnery, E. W., L. B. Kahn, R. L. Reddick, et al. Hemangiopericytoma: a light microscopic and ultrastructural study. Cancer 1981. 47 (5):906–914.
10. Gengler, C. and L. Guillou. Solitary fibrous tumour and hemangiopericytoma: evolution of a concept. Histopathology 2006.48 (1):63–74.
11. Angouridakis N, Zaraboukas T, Vital J, Vital V. Sinonasal hemangiopericytoma of the middle turbinate: a case report and brief review of the literature. B-ENT 2007; 3: 139-143.
12. Higashi K, Nakaya K, Watanabe M, Ikeda R, Suzuki T, Oshima T, Kobayashi T. Glomangiopericytoma of the nasal cavity. Auris Nasus Larynx.2011; 38: 415-417. doi: 10.1016/j.anl.2010.08.009.

DOI: 10.14260/jemds/2014/3995

13. Gillman G, Pavlovich JB. Sinonasal hemangiopericytoma. Otolaryngol Head Neck Surg 2004; 131: 1012-1013.
14. Lester D, Thompson M. Sinonasal tract glomangio-pericytoma (hemangiopericytoma). Ear Nose Throat J 2004; 83: 807
15. Ganly, I., S. G. Patel, H. E. Stambuk, et al. Solitary fibrous tumors of the head and neck: a clinicopathologic and radiologic review. Arch Otolaryngol Head Neck Surg 2006. 132 (5): 517–525.
16. Thompson LD, Miettinen M, Wenig BM. Sinonasal-type hemangiopericytoma: a clinicopathologic and immunophenotypic analysis of 104 cases showing perivascular myoid differentiation. Am J Surg Pathol. 2003; 27: 737-749.
17. Watanabe K, Saito A, Suzuki M, Yamanobe S, Suzuki T. True hemangiopericytoma of the nasal cavity. Arch Pathol Lab Med. 2001; 125: 686-690.

AUTHORS:

1. Somu L.
2. Prasanna Kumar S.
3. Ravikumar A.
4. Raadhika Shree

PARTICULARS OF CONTRIBUTORS:

1. Professor, Department of ENT, Head and Neck Surgery, Sri Ramachandra Medical College, Porur, Chennai.
2. Associate Professor, Department of ENT, Head and Neck Surgery, Sri Ramachandra Medical College, Porur, Chennai.
3. Director and Professor, Department of ENT, Head and Neck Surgery, Sri Ramachandra Medical College, Porur, Chennai.
4. Resident, Department of ENT, Head and Neck Surgery, Sri Ramachandra Medical College, Porur, Chennai.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Prasanna Kumar S,
65/2, East Colony, ICF,
Chennai-38.
Email: sprasannakumar10@gmail.com

Date of Submission: 26/11/2014.
Date of Peer Review: 27/11/2014.
Date of Acceptance: 05/12/2014.
Date of Publishing: 11/12/2014.