

A Case Report on Benign Sinonasal Paraganglioma

Smrity Rupa Borah Dutta¹, Sachender Pal Singh², Aakanksha Rathor³

¹ Assistant Professor; Otorhinolaryngology,
Silchar Medical College & Hospital, Silchar

² PGT; Otorhinolaryngology,
Silchar Medical College & Hospital, Silchar

³ PGT; Otorhinolaryngology,
Silchar Medical College & Hospital, Silchar

Abstract: We report a case of sinonasal paraganglioma presenting with episodes of epistaxis. A 55 year old male presented with a nasal mass. It is an uncommon site of presentation and in an uncommon age group. A high grade of suspicion is required to diagnose sino nasal paraganglioma. However, CT Scan and histopathology helps in early diagnosis and treatment. Surgical excision done with cranialization of frontal sinus with fascia lata graft, followed up for 1 year without any evidence of disease recurrence.

Keywords: Sinonasal; Paraganglioma; Fascia Lata.

1. Introduction

Rarely paraganglioma have been described in areas like the sinonasal tract where there is apparently no paraganglionic tissue and very few cases of definite paraganglioma arising primarily in the nose or paranasal sinuses have been reported, presenting with nasal obstruction, profuse epistaxis and facial swelling. Complete excision of the paraganglioma is normally curative. We report a case of nasal paraganglioma and discuss the diagnosis and therapy. Malignant transformation of benign paraganglioma is rare and transformation of paragangliomas to other types of malignancies is even rarer

2. Case Report

A 55 year old man presented in emergency hours in the department of otorhinlaryngology, silchar medical college & hospital, silchar with history of massive bleeding from nose for last 3 days and a swelling at the root of nose for last 4 months. There had been several episodes of mild intermittent nasal bleeding . There was no complain of nasal obstruction but hyposmia. Immediately anterior nasal pack was given and 1 unit of whole blood was transfused.



Figure 1: Preoperative photo of sinonasal paraganglioma.

2.1. On gross examination, a smooth, firm, non tender, nonpulsatile, diffuse swelling about 4X5 cm size in its greatest dimensions extending from root of nose over its dorsum. The overlying skin was normal with no local rise of temperature.

2.2. ENT examination on anterior rhinoscopy revealed a mass at the roof of the nose bilaterally involving the septum with shift towards right. Nasal endoscopy suggested mass arising from the septum & roof of the nose in the 2nd pass bilaterally.

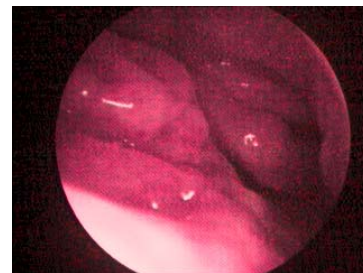


Figure 3: Nasal endoscopy (2nd pass) showing mass in the left nostril arising from the roof & septum of nose.

2.3. Eye examination: Visual acuity, ocular motility was within normal limits, eye lid, conjunctiva, cornea, iris, anterior chamber, lens & pupil of both eye was normal. Fundoscopy was insignificant. Hemoglobin was 8.6gm%, while other laboratory tests were within normal limits. Blood pressure during the pre-operative hospital stay was 150/90 mm Hg (supine position, Left arm).

2.4. Radiological imaging: CECT PNS showed an enhancing expansile soft tissue density mass is noted involving the frontal sinus, bilateral ethmoid sinus, anterosuperior nasal cavity & extending up to subcutaneous plane causing bulging of subcutaneous plane. The mass was causing erosion & remodeling of anterior & posterior walls of frontal sinus, erosion of ethmoidal septae & nasal septum is also noted. Erosion of right lamina papyracea & cribriform plate is noted causing asymmetry of ethmoidal roof. The mass is extending into anterior cranial fossa. Extension into medial & superior aspect of right orbit is also noted.

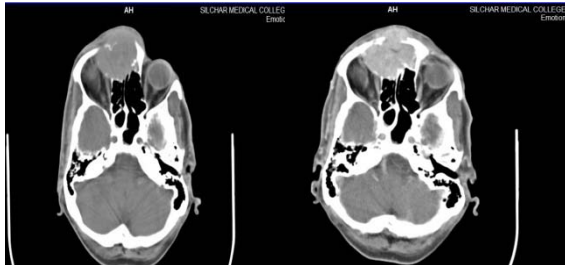


Figure 4.1: CT findings: An enhancing expansile soft tissue density mass is noted involving the frontal sinuses, bilateral anterior ethmoid sinuses, anterosuperior nasal cavity and extending upto subcutaneous plane.

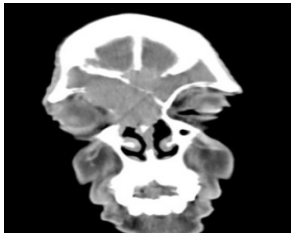


Figure 4.2: Coronal postcontrast image showing extension into anterior cranial fossa.

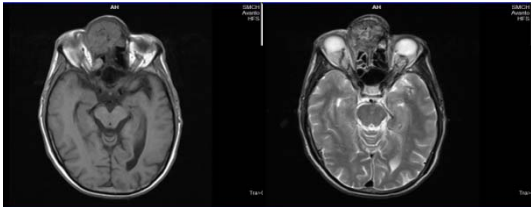


Figure 5.1: MR shows well defined lobulated lesion in fronto-nasal-ethmoidal region which is isointense to brain in T1W and mildly hyperintense on T2W image.



Figure 5.2: Sagittal MR image shows extraaxial extension of lesion into anterior cranial fossa .

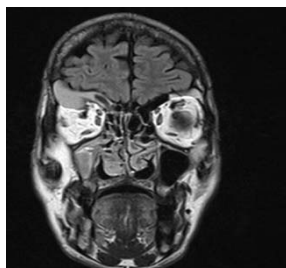


Figure 5.3: FLAIR image show the lesion to be hyperintense with extension into superior aspect of orbit.

3. Operative Details

Under all aseptic & antiseptic conditions general anaesthesia was given. Lumbar drainage catheter was put in I3-I4 space in order to prevent post operative rise of intracranial tension.

Bicoronal flap was raised & tumour was assessed & removed along with surrounding mucosa. Posterior wall of frontal sinus was also removed & tumor was found to be restricted in extradural space. Dura was intact. Estimated blood loss intraoperatively was around 1500ml intraoperatively patient was transfused with 3 units of whole blood & 2000ml of crystalloids.

Cranialization of frontal sinus was done with latissimus dorsi flap. No intraoperative or postoperative complications were found. Lumbar drainage catheter removed after 48 hours when nasal pack was removed after confirming no elevations in the intracranial pressure over 2 days & no CSF leakage.

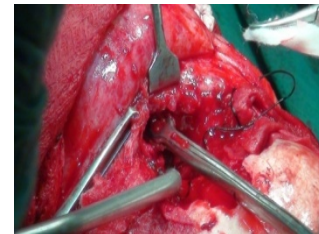


Figure 6.1: Intra operative photo showing removal of tumor.

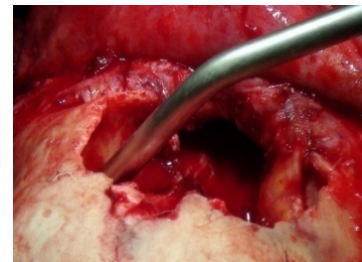


Figure 6.2: Intraoperative photo after the complete removal of tumor.



Figure 6.3: Intraoperative photo showing fascial lata graft.

4. Histopathology Report

A histopathological finding shows it to be a paraganglioma.

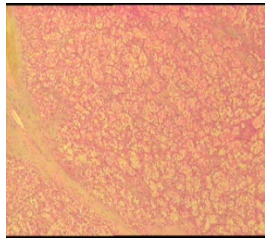


Figure 7.1: Histopathological slide (4X magnification) showing capsule & zellabalen pattern

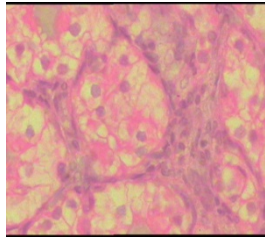


Figure 7.2: Histopathological slide (40X magnification) showing zellabalen pattern

5. Discussion

Paraganglionic chemoreceptor cells of neural crest origin give rise to benign, slow growing but locally invasive tumour known as paraganglioma [1].

Almost half of these occur in the temporal bone, arising from either the cochlear promontory (i.e. typanicum) or the jugular blub (i.e. jugulare), nearly 1/3rd in the carotid body, nearly 1/8th in the region of the high cervical vagus and the rest at various sites of the head and neck[2]. The most common site of occurrence being adrenal glands. In the head and neck area, common sites of occurrence are the carotid body, orbit, larynx, and the nasopharynx, but paragangliomas are rare in the nasal cavity and paranasal sinus.

In the nasal cavity, the middle turbinate, lateral nasal wall and superior nasal vault are the most common sites. In paranasal sinuses, the ethmoid sinus is the most common site of occurrence [4, 5, 6]. Nearly 10 percent of paragangliomas are malignant [7].

In approximately 10 percent of patients, tumours are multifocal and up to 5 percent of tumours secrete catecholamines. [3]

Common symptoms include recurrent epistaxis, nasal obstruction and frontal headache. The clinical presentation depends on the localisation of the tumour. In this case, the patient presented with epistaxis, facial swelling & hyposmia. Hyposmia is probably due to mechanical obstruction by the lesion.

They may also be associated with some syndromes such as multiple endocrine neoplasia type 2b (MEN IIB), von Hippel–Lindau disease, neurofibromatosis types I [8]. In this case, we did not have any syndromic involvement. It is estimated that about 10–50% of paraganglioma cases are familial (autosomal

dominant) [1]. The genes for the familial paraganglioma have been recently identified at the 11q23 locus [9]. Also, 4–19% of all head and neck paragangliomas have been reported to be malignant [10].

The presence of metastasis is the only definite criteria for malignancy as there are no reliable histopathological criteria to distinguish between benign and malignant paragangliomas [1,11] and since the lesions are almost impossible to remove completely, postoperative radiotherapy is then mandatory [3].

In our case, we could not demonstrate any evidence of metastasis to the regional nodes or distant organs. He has been followed up for 12 months, and no additional symptoms or signs indicating recurrence have been identified.

6. Conclusion

Rarely, are the paragangliomas of the sinonasal region reported in the literature. Benign paraganglioma, may occasionally, both clinically and radiologically resemble malignant sinonasal tumour so a high grade of clinical suspicion is required to diagnose such a rare & curable tumor. It may be provisionally diagnosed in any patient with nasal mass associated with severe epistaxis. To conclude, histopathology is the spine for definitive diagnosis.

References

- [1] Pellitteri PK, Rinaldo A, Myssiorek D, Gary JC, Bradley PJ, Devaney KO et al (2004) Paragangliomas of the head and neck. *Oral Oncol* 40:563–575.
- [2] Zak FG, Lawson W. *The paraganglionic chemoreceptor system. Physiology, pathology and clinical medicine*. New York: Springer Verlag. 1982.
- [3] Scott Brown's Otorhinolaryngology, Head & Neck Surgery; 7th edition 2008.
- [4] Sharma HS, Madhavan M, Othman NH, Muhamad M, Abdullah JM. Malignant paraganglioma of frontoethmoidal region. *Auris Nasus larynx* 1999;26:487–93.
- [5] Welkoborsky HJ, Gosepath J, Jacob R, Mann WJ, Amedee RG. Biologic characteristics of paragangliomas of the nasal cavity and paranasal sinuses. *Am J Rhinol* 2000;14:419–26.
- [6] Mevio E, bignami M, Luinetti O, Villani L. Nasal paraganglioma: a case report. *Acta Oto-rhinolaryngologica Belg* 2001; 55:247–9.
- [7] Conley JJ. The carotid body tumour: A review of 23 cases. *Archives of Otolaryngology*. 1965;81 : 187-93.
- [8] Bijlenga P, Dulguerov P, Richter M, de Tribolet N (2004) Nasopharynx paraganglioma with extension in the clivus. *Acta Neurochir (Wien)* 146:1355–1359.
- [9] Baysal BE, Van Schothorst EM, Farr JE, Grashof P, Myssiorek D, Rubinstein WS et al (1999) Repositioning the hereditary paraganglioma critical region on chromosome band 11q23. *Hum Genet* 104:219–225.
- [10] Kuhn JA, Aronoff BL (1989) Nasal and nasopharyngeal paraganglioma. *J Surg Oncol* 40:38–45.
- [11] Deb P, Sharma MC, Gaikwad S, Gupta A, Mehta VS, Sarkar C (2005) Cerebellopontine angle paraganglioma—report of a case and review of the

literature. J Neuro-oncol 74:65–69Subsidiary, H. Etemad and L. S, Sulude (eds.), Croom-Helm, London, 1986. (book chapter style)

- [12] K. Deb, S. Agrawal, A. Pratab, T. Meyarivan, “A Fast Elitist Non-dominated Sorting Genetic Algorithms for Multiobjective Optimization: NSGA II,” KanGAL report 200001, Indian Institute of Technology, Kanpur, India, 2000. (technical report style)
- [13] J. Gerald, "Sega Ends Production of Dreamcast," vnunet.com, para. 2, Jan. 31, 2001. [Online]. Available: <http://nl1.vnunet.com/news/1116995>. [Accessed: Sept. 12, 2004]. (General Internet site)

Author Profile



Dr. Smrity Rupa Borah Dutta completed her M.B.B.S and M.S degrees from Assam Medical College, Dibrugarh, Assam in the years 2000 and 2004 respectively. Presently she is the Assistant Professor in the Department of Otorhinolaryngology, Silchar Medical College, Assam, India.