

## CASE REPORT

# Sinonasal Paraganglioma: A Case Report and Review of Literature

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## ABSTRACT

A rare case of sinonasal paraganglioma is described. A 40-year-old female patient presented with 2 years history of unilateral nasal obstruction and bleeding. CT scan demonstrated an expansile enhancing mass involving bilateral ethmoids, right nasal cavity and right maxillary sinus. Histopathological diagnosis was neuroendocrine tumor with possibility of paraganglioma. A subtotal maxillectomy with excision of mass performed. Primary nonchromaffin paraganglioma of nose and paranasal sinus is a very rare tumor, these lesion pose diagnostic challenge to clinicians and pathologist. Only twenty five cases are reported in review of literature. Surgical excision is the mainstay of treatment.

**Keywords:** Paraganglioma, Sinonasal, Excision, Endoscopic.

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## INTRODUCTION

Paragangliomas are tumors of the autonomic nervous system, arising from paraganglionic tissue. Paragangliomas of the head and neck region are very rare.<sup>1</sup> Head and neck paragangliomas are commonly nonfunctional, non-chromaffin tumors. They are most commonly seen in carotid body, temporal bone and along the vagus nerve. Sinonasal paragangliomas are extremely rare. Only twenty five cases have been reported worldwide.<sup>2</sup> We report a case of sinonasal paraganglioma in an adult female patient along with the review of literature.

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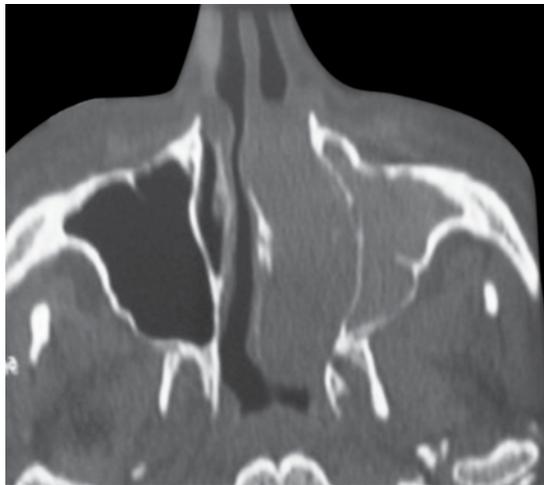
A 40 years old female presented to ENT-OPD with complaints of right side nasal obstruction and occasional epistaxis for last 2 years duration, which was insidious in onset and gradually progressive. Patient was treated by her general practitioner with antibiotics and antihistamine during the course of treatment but showed no improvement. On external examination there was expansion of nasal pyramid. On anterior rhinoscopy a pinkish mass was seen filling the entire right nasal cavity, which was soft inconsistency, non friable, insensitive and bled on touch. It was found attached to the right lateral nasal wall.

General physical examination of patient was normal. There was no history of palpitation, diaphoresis and headache. Patient was normotensive, nondiabetic. Routine blood and urine examination was within normal limit. No similar complaint was found in any of the other family member.

Nasal endoscopic biopsy was done which was suggestive of neuroendocrine tumor with possibility of paraganglioma.

Contrast enhanced computed tomography of paranasalsinus (Fig. 1) showed an expansile mass with patchy enhancement which involved bilateral ethmoids, right nasal cavity, right maxillary sinus and abutting the anterior skull base. Also there was lateral bowing of medial wall of right maxillary sinus and mass was seen extending into nasopharynx (Fig. 2). Ultrasound abdomen was done during the course of investigation, revealed a mass on lower pole of left kidney, for which nephrectomy was done in department of surgery and biopsy was suggestive of clear cell variety of renal cell carcinoma.

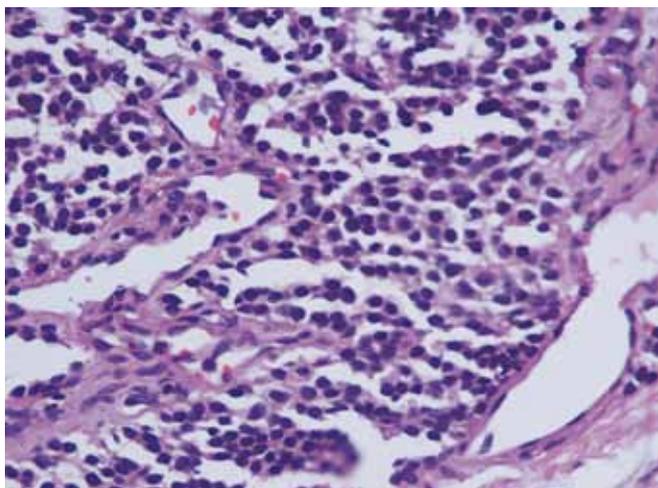
Patient was then planned for surgery. Right side subtotal maxillectomy was done under general anesthesia by lateral rhinotomy approach. Intraoperative a pink colored exophytic mass arising from maxillary and ethmoid sinuses, extending into nasopharynx was visualized. Maxillary sinus was filled with tumor tissue and thick retained secretions. Medial wall of maxillary sinus was papery thin and was removed along with the anterolateral and a part of posterolateral wall. Patient's blood pressure was stable during the procedure and no pressure fluctuation were noted, cardiac rhythm during the surgery was regular and normal. Nasal packing was done at the end of the procedure after complete hemostasis,



**Fig. 1:** Contrast enhanced coronal CT scan of paranasal sinuses showing expansile enhancing mass in right nasal cavity, ethmoids, maxillary antrum and thinning of medial wall of maxilla



**Fig. 2:** Contrast enhanced axial CT scan of paranasal sinuses with tumor extension to maxillary sinus and nasopharynx



**Fig. 3:** Microphotograph (H and E stain, magnification,  $\times 400$ ) of the case showing polygonal tumor cells present in sheets with thin walled blood vessels in between tumor cells

which was removed on third postoperative day. Patient's postoperative period was uneventful.

Histopathology report of surgical specimen, confirmed the lesion as a paraganglioma of right Nasal cavity and paranasal sinuses (Fig. 3).

## DISCUSSION

Paraganglioma is a rare tumor. Kohn first described paraganglioma as the 'organs of zuckerandl' which were disk like collection of tissue in the adventitia of the aorta. Paragangliomas are found throughout the body, usually closely associated with neural and vascular structures. They were classified into four main group by Glenner and Grimly<sup>3</sup>: (1) branchiomic, (2) intravagal, (3) aortic sympathetic and (4) visceral-autonomic. Paraganglioma of head and neck are more closely associated with the parasympathetic system and are of nonchromaffin type. Carotid paraganglioma are the most common, followed by jugulotympanic, vagal and

laryngeal in descending order of frequency. While rarely found in the orbit, pterygopalatine fossa, thyroid, nasopharynx and sphenoid and maxillary sinuses.<sup>4</sup> A review of literature showed only twenty five cases of nonchromaffin paragangliomas of nasal cavity and paranasal sinuses worldwide<sup>2</sup> and only twelve cases of nasal cavity mucosa involvement primarily.<sup>5</sup>

True incidence ranges from 1:30,000<sup>6</sup> to 1:500,000.<sup>7</sup> There is no obvious age and sex predominance, but a large series of benign paragangliomas reported a mean age of 47 years. The majority of paraganglioma are benign, slow growing but locally invasive and destructive lesions.<sup>8</sup> Malignancy in head and neck paragangliomas is rare.<sup>5</sup>

Histopathologically paragangliomas are composed of two distinct types of cell. Type 1 cells have the capacity for the synthesis and storage of catecholamine and characterized by a rich concentration of cytoplasmic organelles and hormone-containing granules. Type 2 cells are sustentacular cells and are similar to Schwann cells morphologically. Sustentacular cells are typical structural elements of the normal adrenal and extra-adrenal paraganglioma system. S-100 protein is present in Schwann cells, sustentacular cells and glial cells.

Antibody to S-100 protein is used in immunostaining of sustentacular cells. Malignant paraganglioma are completely devoid of S-100 positive sustentacular cells although some benign tumors may also be devoid of sustentacular cells.<sup>9</sup> Nonsecretory paragangliomas consist of focal collections of neuroepithelial chief cells which are arranged in well-defined nests, which have the classic alveolar or so-called 'zellballen' pattern. They are encircled by sustentacular cells. Immunohistochemistry can be of help not only in diagnosis but may also assist with regard to prognosis.<sup>2</sup>

Management of paragangliomas is controversial. Surgery is the mainstay of treatment with maximal consid-

ration given to adequate excision to achieve tumor free margins. Furthermore, because of the aggressive nature of these tumors, regardless of the surgical approach, long-term follow-up is recommended. Recurrence is a common following excision of tumor. This case was followed up regularly and no recurrence was observed in 24 months period. Radiotherapy has been kept as an option for any future recurrence.

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