

Case Report

Paraganglioma of nose: a common tumor, rare site

Sanjay Khandekar, Pratik Chide*, Nilima Lodha, Jayawant Mahadani, Snehalata Hingway,
Vikas Yedshikar, Keshav Pagrut, Hitendra Khandekar, Vijay Kunghadkar

Department of Pathology, SVNGMC Yavatmal, Maharashtra, India

Received: 01 May 2016

Accepted: 09 May 2016

***Correspondence:**

Dr. Pratik Chide,

E-mail: pratikchide@gmail.com

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ABSTRACT

Paragangliomas are neuroendocrine tumors, arising from paraganglionic tissue. They most commonly originate in the adrenal gland and are known as pheochromocytoma. However 5-10% of these tumors are extra-adrenal and may appear in the neck and pelvic region along the sympathetic nervous system.¹ In the head and neck, the most common sites of origin of this neoplasm are the carotid body, the jugular bulb and the vagal body. Paragangliomas arising from the nose are very rare. We here report a rare case of paraganglioma of the nose diagnosed on histopathology.

Keywords: Paraganglioma, Neuroendocrine tumors, Nasal cavity, Adrenal gland neoplasms

INTRODUCTION

Paraganglioma is a neuroendocrine neoplasm arising from paraganglionic tissue of the autonomic nervous system. Paraganglions have two types of cells: Type I, which contain catecholamine granules and Type II, which are supportive cells similar to Schwann cells surrounding type I cells.¹ Paraganglions are broadly distributed in the human body, found in the lungs, heart, mediastinum, gastrointestinal tract, retroperitoneal region and bladder. In the head and neck, they were found in the orbit, larynx, nasopharynx, thyroid gland, vagus, mediastinum, lung, duodenum, aortic region (organ of Zuckerkandl), retroperitoneal region, and bladder, tongue, hypophysis, pineal gland.¹ The term paraganglioma is currently suggested by several authors for all tumors derived from paraganglion tissue regardless of the result of chromaffin dye staining. This is because that less than 1% of paragangliomas exhibit only endocrine activity.² In this study we report a rare case of nasal paraganglioma in a 15-year-old male patient.

CASE REPORT

A 15-year-old male complained of unilateral nasal obstruction and nasal bleeding for 2 months and was hospitalized for 5 days to control the bleeding. In the ENT examination, there was a mass in the right nostril that shifted the wall of the maxillary sinus sideways and pushed the nasal septum to the opposite side. The left nostril was entirely blocked. Contrast-enhanced computed tomography (CT) showed a 3.2 cm X 2 cm mass in the axial position with an expansive character completely occupying the right nasal cavity from the middle nasal turbinate to the nasopharynx.

A total resection of the mass was done. On gross examination, the specimen was in the form of multiple fragments, aggregating to a size of 3x2 cm with a grayish to pinkish color and firm to hard consistency. On histological examination, the tumor mass shows tumor cells arranged in cords and an alveolar pattern (zellballen pattern) which are separated by highly vascularized stroma. Individual tumor cells are round to polyhedral in shape with round vesicular nuclei and eosinophilic cytoplasm. Mitosis was absent. With the basic idea of

paraganglioma to rule out glomus tumor and haemangiopericytoma, tissue was sent for immunohistochemistry for confirmation of diagnosis. As on immunohistochemistry, tumor cells were positive for synaptophysin and focally for S100 protein, so the diagnosis of paraganglioma of nose was confirmed. The tumor cells are immunonegative for chromogranin/EMA/Pax8/CD10/TFE3.

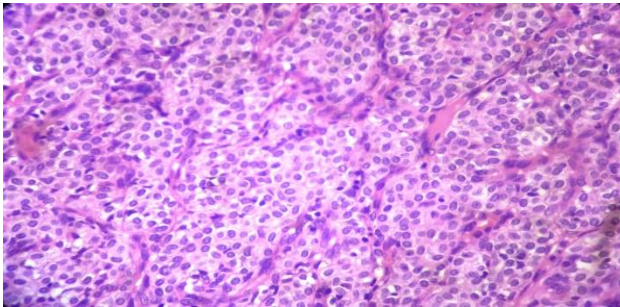


Figure 1: Tumor cells arranged in alveolar pattern (zellballen pattern).

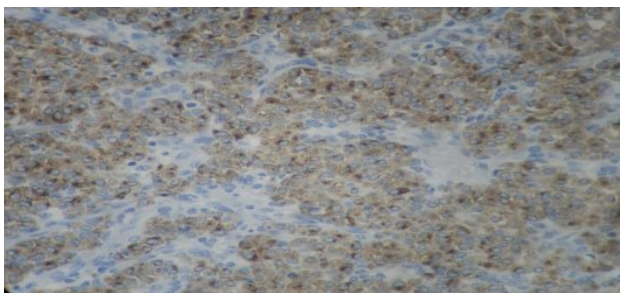


Figure 2: Tumor cells are diffusely positive for synaptophysin.

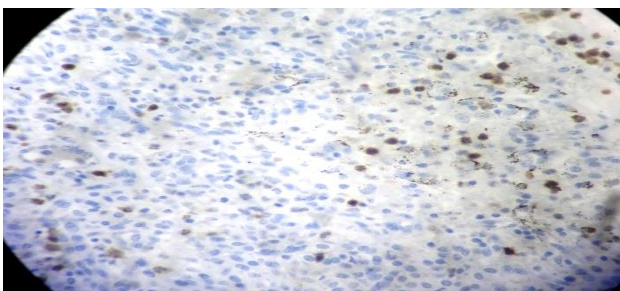


Figure 3: Tumor cells are focally positive for S-100.

DISCUSSION

Nasal paragangliomas are very rare; until 2002, there were only 27 cases reported in the literature. According to a study comprising of 73 cases of head and neck paragangliomas, only 3 were of nasal and paranasal types³The tumor occurs in a wide range of patients aged between 8 and 89 years, with middle-aged females being most affected.⁴ In this case report patient was 14 year old male.

The nasal cavity is more affected by the paraganglioma than the sinuses; most paragangliomas originate in the lateral nasal wall and middle turbinate. Regarding the location of this tumor in the nasal cavity, it is worth mentioning that in the first 3 cases described in the literature, the source of the tumor was the nasal septum.⁵ Tumors originating from the inferior turbinate and the posterior portion of the choana have also been described.⁶ In our case tumor was occupying the right nasal cavity from the foyer to the nasopharynx.

Paragangliomas are slow-growing, expansive, multicentric tumors. Approximately 3% of these tumors are extra-adrenal, and up to 26% of patients with this tumor have a positive family history, illustrating the existence of genetic predisposition⁷ in our case there was no positive family history.

The clinical presentation is usually characterized by nasal obstruction, repeated and profuse bleeding, a runny nose, facial swelling, and ipsilateral pain⁸ In this case patient had complained of unilateral nasal obstruction and nasal bleeding. They may also be associated with some syndromes such as multiple endocrine neoplasia type 2b (MEN IIB), von Hippel-Lindau disease, neurofibromatosis Type I. In this case, we did not have any symptoms associated with syndrome.

Macroscopic examination of the paraganglioma revealed firm to hard in consistency with grayish or rosy in colour and apparently encapsulated lesions.^{8,9} In this case, grossly tumor mass was in multiple fragments with grayish to pinkish in colour and firm to hard in consistency.

The histopathological differential diagnosis also includes olfactory neuroblastomas, meningioma, haemangiopericytoma, angiosarcoma, spindle cell sarcoma and poorly differentiated squamous cell carcinoma. Both paraganglioma and olfactory neurofibroma can be recognised among these tumours on the bases of their neuroendocrine phenotype (with NSE-, synaptophysin-and chromogranin positive chief cells and S-100 protein-positive sustentacular cells). Olfactory neuroblastoma can be differentiated from paraganglioma because its morphology is quite distinctive (presence of large tumour nest, neuropil and rosettes).

Immunohistochemistry can be of help not only in diagnosis but may also assist with regard to prognosis.¹² Achilles et al assessed the value of immune histochemical staining for S-100 protein in the diagnosis and prognosis of paraganglioma and observed that malignant paraganglioma were completely devoid of S-100 positive sustentacular cells; however, a small proportion of benign tumours were also lacking in sustentacular cells, thus making it difficult to reach an unequivocal conclusion about the reliability of this finding. Nonetheless positivity for S-100 and synaptophysin of tumour cells in the present case (Figure 2).

Malignant head and neck paragangliomas show an incidence of 4 to 19 %.¹⁰ Metastases are rare and are found in 9% of the cases.¹¹ Literature reports show that metastases tend to involve lymph nodes, lungs and bones.³ It is widely accepted that malignancy potential of paragangliomas cannot be estimated by lesions' histological aspect only.³ Not only histological findings suggestive of potential malignant behavior such as mitotic figures, nest necrosis and vascular invasion should be considered, but also an accurate assessment of paragangliomas presenting uncommon infiltrating growth or recurrences¹¹ should be performed. Such neoplasias should be submitted to aggressive surgical resections, once they are strongly characteristic of malignant behavior.¹⁰

CONCLUSION

The appearance of these lesions in the nasal cavity and paranasal sinuses is extremely rare. Clinically, they cause obstruction and nasal bleeding, and they can be mistaken for other benign and malignant lesions. They are histologically and immunologically different from glomus tumors, but these lesions should not be confused with those even though they share the characteristic of great vascularity. The treatment should be surgical, as other therapies are palliative and surgery tends to be radical in terms of complete elimination and prevention of recurrence.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Khandekar S, Chide P, Lodha N, Mahadani J, Hingway S, et al. Paraganglioma of nose: a common tumor, rare site. *Int J Res Med Sci* 2016;4:2475-7.