

## Sinonasal paraganglioma: a case report

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### Abstract

**Objective** To highlight the importance of a high index of suspicion and meticulous evaluation of a patient with bleeding sinonasal mass in the diagnosis of sinonasal paraganglioma.

**Method** Case report of a 39-year-old female who presented with a bleeding lobulated left nasal mass necessitating multiple blood transfusions is presented.

**Result** Diagnostic investigations revealed that the patient was HIV-positive (ELISA and Western blot) and a tumour histology of paraganglioma. She had a CD4<sup>+</sup> count of 487 cells/mm<sup>3</sup>. The urinary vanillylmandelic acid assay and

echocardiography were normal. Subsequently, she had complete tumour excision through a medial maxillectomy and has remained without a recurrence 12 months after.

**Conclusion** To our knowledge, this is the first report in the world literature of sinonasal paraganglioma in a HIV-positive patient and the first reported case of sinonasal paraganglioma in Africa. It is, however, not clear from this report if the patient's HIV status preceded her development of the paraganglioma or not.

**Keywords** Epistaxis · Neuroendocrine tumours · Sinonasal paraganglioma

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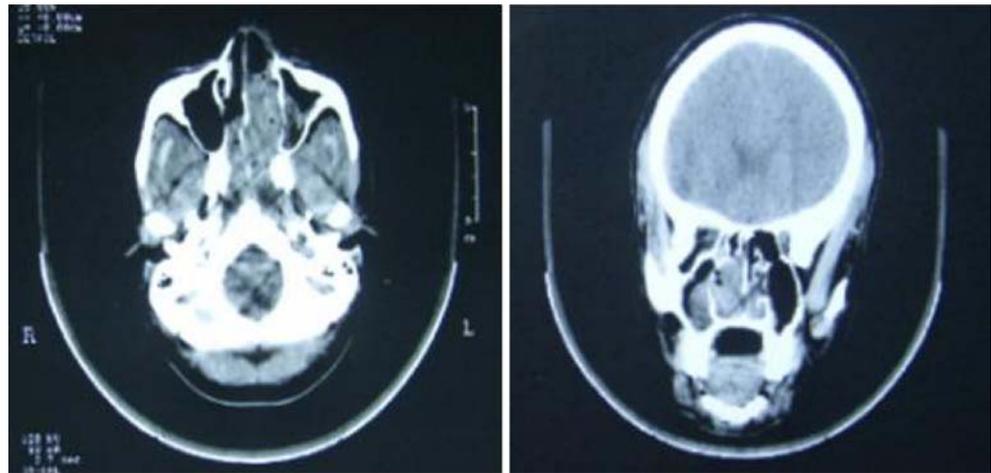
### Introduction

Paragangliomas of the head and neck are mostly benign, slowly growing neuroendocrine tumours which arise from the extra-adrenal paraganglia of the autonomic nervous system [1]. They are distributed along the course of the major vasculature and those that occur in the head and neck region arise from the parasympathetic nervous system [1, 2]. Paragangliomas of the head and neck region are very rare and constitute 0.01–0.6% of all tumours in the head and neck region of which approximately 80% are located either in the carotid body or in the jugular bulb [3].

Paraganglioma of the nose and paranasal sinuses are very uncommon [4]. It has been suggested that paraganglioma originate from ganglionic tissues closely related to the arteries and nerves in the pterygopalatine fossa [5]. Majority of the earlier reported cases of the tumour were in the ethmoidal region and the middle concha [6].

It is the purpose of this communication to highlight this first case of nasomaxillary paraganglioma in an adult female in Africa; MEDLINE search have revealed no previous case.

**Fig. 1** Computed tomography scan of the patient's paranasal sinuses



### Case report

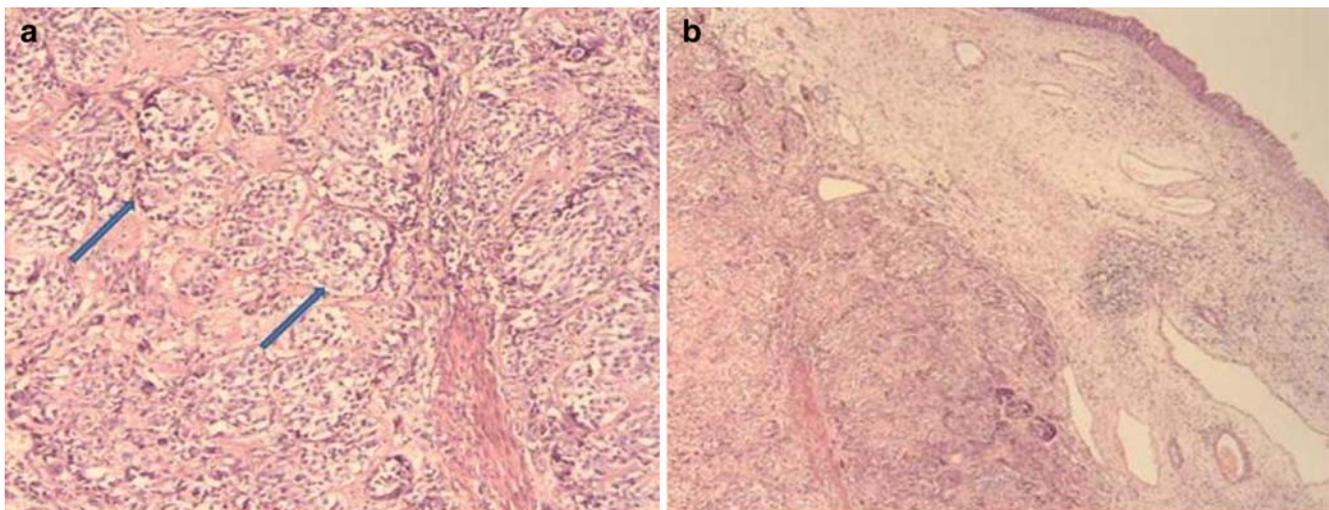
A 39-year-old lady presented in the ENT clinic of our centre with 15 months history of spontaneous, recurrent epistaxis associated with progressive left nasal growth and blockage. There was associated snoring, hyponasal speech, mouth breathing, feeling of aural blockage and impaired hearing on the left ear. She also had easy fatigability and recurrent fainting attacks which were noted 4 months before presentation. There was no history of cheek or neck swelling, loose or lost tooth, cough, yellowness of the eye, abdominal swelling and bone pain. She had had multiple blood transfusions from other private health facilities before referral to us. She had no family history of similar illness.

Examination revealed an afebrile young female with noisy breathing, hyponasal speech and was pale. She had a pulse of 125 bpm and a blood pressure of 70/50 mmHg. The external nasal pyramid appeared normal; however, the

left nasal cavity was completely obliterated by a lobulated fleshy haemorrhagic mass covered by sanguineous mucoid secretions. The mass was firm, sensitive to touch, bled easily on contact and appeared to arise from the left lateral nasal wall. The septum was slightly displaced to the contralateral side. The throat examination only revealed postnasal mucoid discharge mixed with altered blood. Auroscopy revealed a retracted dull left tympanic membrane. The neck and eyes appeared normal.

At the time of examination, she had profuse bleeding from the left nasal cavity for which gauze pack was introduced into the nasal cavity to serve as a tamponade. She was resuscitated with crystalloids. An urgent packed cell volume done was 13% and she was subsequently transfused with fresh whole blood after blood sample had been taken for HIV screening with patient's consent.

The computed tomography scan of the paranasal sinuses done after the patient had become haemodynamically stable



**Fig. 2** **a** Organoid clusters of polygonal- to spindle-shaped cells characteristic of zellballen arrangement (haematoxylin–eosin,  $\times 40$ ). **b** A section of sinonasal paraganglioma showing overlying respiratory epithelium (haematoxylin–eosin,  $\times 40$ )

revealed an expanded left nasal cavity with soft tissue density lesion extending into the nasopharynx and the left maxillary antrum (Fig. 1).

A pernasal biopsy of the left nasal mass was done afterwards and histology revealed an infiltrative neoplasm comprising of organoid clusters of polygonal- to spindle-shaped cells characteristic of zellballen arrangement with a vascularised stroma areas of fibromyxoid changes and focal hyalinisation. Features were consistent with a paraganglioma (Fig. 2a,b).

The vanillylmandelic acid and echocardiography were normal, whilst the retroviral test was positive (ELISA and Western blot). The CD4<sup>+</sup> count was 487 cells/mm<sup>3</sup>.

She subsequently had a left medial maxillectomy through a lateral rhinotomy approach. The operative findings were a yellowish, firm, left intranasal mass which extended into the nasopharynx and left maxillary antrum via its dehiscent medial wall. A complete resection of this tumour was achieved without any complication intra-operatively.

She had a sustained post-operative clinical improvement and was discharged to the outpatient clinic for follow-up. The final histologic diagnosis was consistent with the initial biopsy report. Monthly fibre optic nasal endoscopy, three-monthly plain radiography of the chest and routine general clinical evaluation of the patient have not revealed any evidence of metastasis or recurrence a year after.

## Discussion

Paragangliomas of the head and neck are usually benign tumours which are slow-growing but locally invasive [1]. The clinical presentation depends on the localisation of the tumour. In this case, the patient presented with recurrent profuse epistaxis, nasal blockage and fainting attack, which necessitated repeated blood transfusion from peripheral hospitals with a high risk of HIV infection [7]. It is, however, not clear from this report if the patient's HIV status preceded her development of the paraganglioma or not. There is no definite association of paraganglioma with HIV infection documented in the literature, and this occurrence may likely be coincidental.

The majority of paragangliomas are solitary lesions, but they may present as multiple tumours with or without secretory characteristics in approximately 10% of the cases. They may also be associated with some syndromes such as multiple endocrine neoplasia type 2b (MEN IIB), von Hippel–Lindau disease, neurofibromatosis type I [2]. 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 [8]. Also, 4–19% of all head and neck

paragangliomas has been reported to be malignant [9]. 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, 10]. In our case, we could not demonstrate any evidence of metastasis to the regional nodes or distant organs and familial trait.

Paragangliomas have a characteristic histological architecture; the individual tumour cells are arranged in distinctive cell balls (zellballen) which consist of type I (chief cells) and type II (sustentacular cells) [1]. At light microscopy, paragangliomas may be extremely difficult to differentiate from sinonasal neuroendocrine carcinomas because both demonstrate neurosecretory granules by electron microscopy. However, immunohistochemical staining for keratin may be helpful in this setting being positive in neuroendocrine carcinoma. 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 (large tumour nest, neuropil and rosettes are present) [11].

The main management modalities of paragangliomas are surgery and radiation therapy. The aim of surgery is complete tumour excision because these tumours may be destructive locally and even more imperative in cases of sinonasal paraganglioma which have higher incidences of malignant behaviour [12]. However, limited tumours can be resected by endoscopic surgery, as was the case of Lecanu et al. [13]. Our patient had complete tumour excision via a medial maxillectomy. This option was adopted because of the large tumour size which filled and splayed the entire left nasal cavity enabling a better visualisation of the tumour extent and entire nasal cavity/nasopharynx. The goal of conventional fractionated, external beam radiation therapy is long-term tumour control [14]. No radiotherapy was given because there was no evidence of malignant transformation in our case. She has since been followed-up for 12 months, and no additional symptoms or signs indicating recurrence have been identified.

In conclusion, paragangliomas of the sinonasal (nasomaxillary) region are extremely rare. Clinical and radiological appearances may resemble malignant sinonasal tumour. It should be considered in any patient with nasal mass associated with severe epistaxis. Histopathological and immunohistochemical diagnosis is essential for a definitive diagnosis.

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