CASE REPORTS

Paraganglioma of maxillary sinus associated with Addison's disease mimicking a vascular tumour: A case report

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ABSTRACT

Paraganglioma of the nasal and paranasal sinuses are quite rare neuroendocrine tumours, usually hormonally inactive, presenting as nasal polyps with or without epistaxis. We present a rare case of a 23-year old male, with Addison's disease, who was referred from endocrinology department with a complaint of right nasal blockage associated with recurrent epistaxis. A provisional clinical diagnosis of vascular tumour was made, computed tomography (CT) and magnetic resonant imaging (MRI) suggested hemangioma/inverted papilloma. However, biopsy reported a highly vascular tissue mimicking angiofibroma, further immunostaining studies with Synaptophysin and S-100 confirmed the diagnosis of paraganglioma. This rare tumour in the paranasal sinuses may mimic vascular tumour and can only be confirmed with histopathological immunostains studies. According to our knowledge, this is first case reported to be associated with Addison's disease in the English literature.

Key Words: Sinunasal paraganglioma, Management, Chemodectoma, Addison's disease, Cushing's syndrome

1. INTRODUCTION

Paraganglioma are slow growing tumours of autonomous nervous system and account for 0.5% of all head and neck tumours, they commonly arise from carotid body, jugular bulb and vagal and rarely from other regions including sinonasal, orbit, larynx.[1] Originating from the neural crest, these vascular tumors may be hormonally active producing adrenocorticotropic hormone (ACTH), parathyroid-related peptide, antidiuretic hormone, intestinal vasoactive peptide or growth hormone, manifesting clinically as Cushing's syndrome, hypercalcemia, inappropriate secretion of antidiuretic hormone syndrome or acromegaly respectively, depending on their neurosecretory granules content.[2]

The reported sinonasal paraganglioma are < 5% of all head and neck paraganglioma[3] with a predilection for middle age females,[4] they usually present with history of nasal obstruction associated with[5] or without epistaxis[6] and or very rarely as Cushing's syndrome due to ACTH secretion.[7–9] Computed tomography (CT) and magnetic resonant imaging (MRI) are the investigations of choice,[10] however, none has a pathognomonic sign for the disease. Their behavior is variable, ranging from malignant locally aggressive with metastases to simple benign expanding mass. And diagnosis is based on histopathological findings of the classical Zellbellan cell nests and sustentacular cells highlighted by S-100 immunostains. However, atypical morphology may be encountered.[11]

This variability, however, creates a challenge for both treating

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physician and histopathologists alike, especially if there is an association with systemic disease, *i.e.* Cushing's syndrome or Addison's disease, as seen in our case.

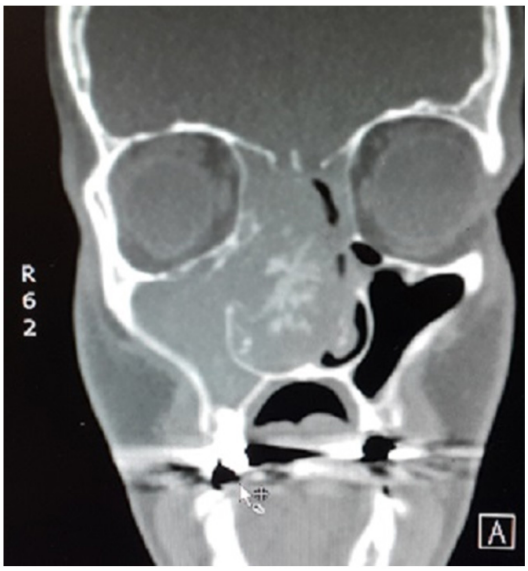


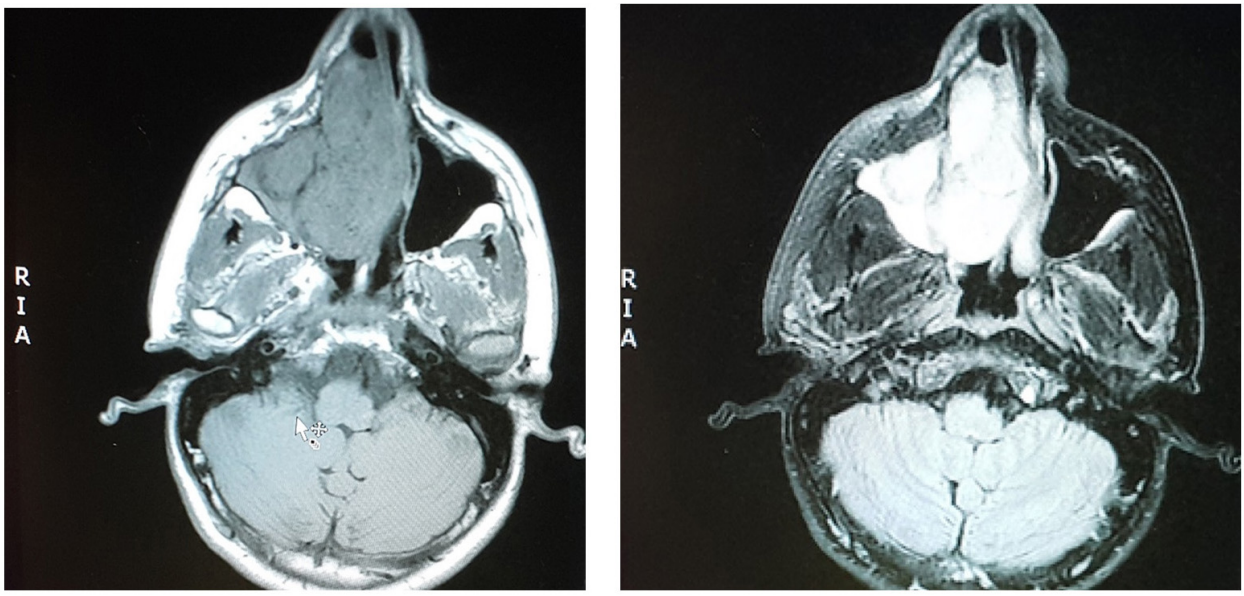
Figure 1. Coronal CT Scan demonstrating a homogenous mass of right maxillary sinus with scalloping of septum

2. Case presentation

A 23-year old male known case of Addison's disease on fludrocortisone referred to our Otorhinolaryngology outpatient department with a complaint of chronic right nasal blockage and recurrent epistaxis. He had multiple hospital admission related to the primary disease.

On presentation laboratory report was: Hemoglobin 12 gm/L, White cell count 2.24, Platelet 224/mm^3, Na 133 mmol/L and K 5.1 mmol/L. And Blood pressure 120/70 and pulse 80/min. Nasal endoscopic examination showed a large polyp in the right nasal cavity which was not actively bleeding. Considering his general condition, the option of a biopsy under local anaesthetic was taken, which was followed by copious bleeding that was controlled by nasal packing.

CT of the paranasal sinuses revealed a homogenous mass occupying right maxillary sinus extending to the right nasal cavity with scalloping of septum, with no evidence of bony erosion (see Figure 1), and MRI showed a hypertense on fluid-attenuated inversion recovery (FLAIR) and T2W1, while it was hypotense on T1 (see Figure 2) with diffuse enhancement in T1W1- Post contrast. The differential diagnosis included, vascular tumour/inverted Papilloma.



A B

Figure 2. A) MRI T1 Demonstrating hypotense mass in right maxillary sinus; B) MRI demonstrating diffuse enhancement with T2W1 post contrast

The initial histopathology reported a highly vascular tissue mimicking angiofibroma, however, further studies with neuron specific enolase (NSE), CD56, synaptophysin and S-100 stain showed sustentacular cells (nerve twigs) confirmed the diagnosis of paraganglioma (see Figure 3).

Surgical option was offered to the patient, however, he opted to be treated elsewhere.

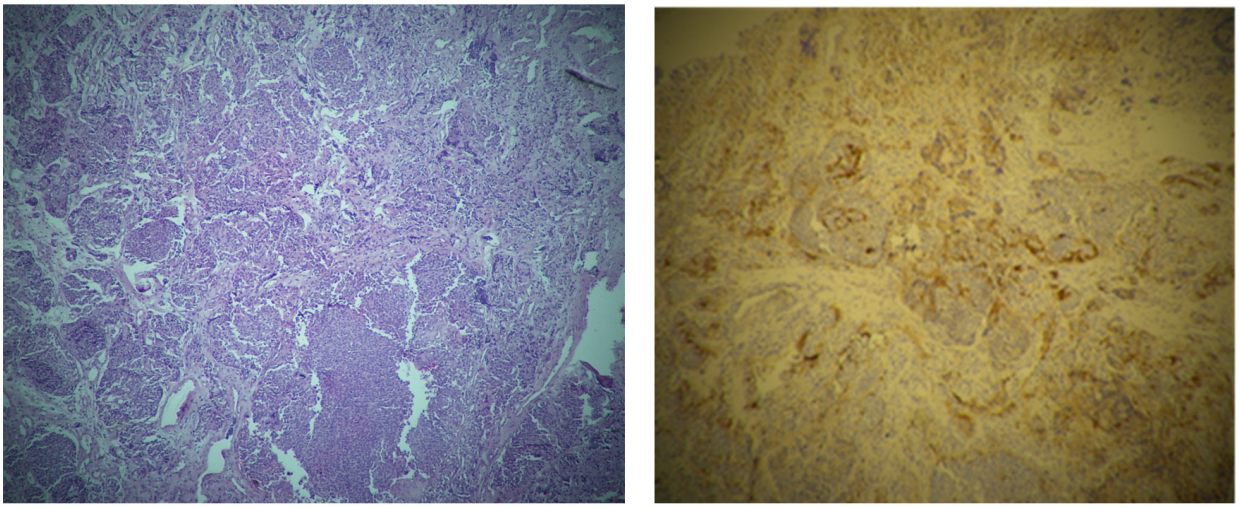


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Figure 3. A) Classical Zellbellan cell nests; B) S-100 stains highlighting sustentacular cells (nerve twigs)

3. Discussion

The clinicopathology of sinonasal paraganglioma in comparison to other head and neck paraganglioma are not quite well understood, arising from middle turbinate, fronto-ethmoid air cells,[4] sphenoid[12] and maxillary sinus,[13] skull base[5] and pterygopalatine fissure,[10] they are rarely hormonally active with only three reported cases,[8,9] moreover they have no named related Paraganglions.

Although these tumors are mainly benign, aggressive malignant clinical behavior with metastasis was noted in some cases where their morphology showed no features of mitosis.[14] This, however, was not shared by others.[15]

Furthermore, radiotherapy that is used as a mode of treatment has been implicated in its malignant transformation.[1]

Diagnostic challenges arises when clinical features as well as imaging modules including CT, MRI and PET are non-specific,[6] and the typical morphology findings mentioned above are atypical, *i.e.* lipoblast-like vacuolated cells.[14]

Our pathology department used Immuno-Histo-Chemistry (IHC) Ventana detection system benchmark XT: In which CD56, Synaptophysin and Neuron Specific Enolase (NSA) were positive while Alpha Smooth Muscle Actin Antibody (ASMA) highlighted a dense vascularity particularly of the lesion area. This might explain the initial pathologist's findings.

The association of sinonasal paraganglioma with Addison's disease, an autoimmune condition, seen in our case and not previously reported, is probably un-related. Whereas, its association with Cushing's syndrome[7] and AIDS[17] in other reports, can be explained by the ectopic hormonal affect and blood transfusion respectively.

In conclusion, sinonasal paraganglioma, indeed is a rare entity, and has variable presentations, little is known about its Pathiophysiology. Its diagnosis remains to be a challenge, listing it among the differential diagnosis may help future management plan strategies.

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Conflicts of Interest Disclosure

The authors declare that there is no conflict of interest statement.

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