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Clinical Record

Primary carcinoid tumour of nasal septum

T Galm, N Turner

Abstract

Objective: We present the first reported case of primary carcinoid tumour of the nasal septum.

Method: Case report of our experience of a carcinoid tumour of the nasal septum. We discuss our clinical, radiological and pathological findings.

Result: An 83-year-old woman presented with a history of left-sided nasal blockage. Clinical examination showed a unilateral, left-sided nasal polyp. Further imaging and histological analysis confirmed this to be a carcinoid tumour. Carcinoid tumours outside the gastrointestinal tract are rare. There have been reports of carcinoid tumours in the head and neck region, but no published cases occurring in the nasal septum. Our management involved wide surgical resection with regular follow up to monitor for recurrence and for the development of carcinoid syndrome. Four years from initial presentation, the patient remained free of the primary tumour and had displayed no signs or symptoms suggestive of carcinoid syndrome.

Conclusion: To the authors' best knowledge, and after searching the world literature, the presented case represents the first report of primary carcinoid tumour of the nasal septum. Despite its rarity, this tumour should be considered as part of the differential diagnosis, as timely recognition and intervention are critical for successful treatment.

Key words: Nasal Septum; Carcinoid Tumour; Histology

Introduction

Tumours of the nasal septum are rare. There have been reported cases of carcinoid tumour in the head and neck region, involving the larynx,[1] parotid[2] and middle ear.[3] However, there have been no reported cases of primary carcinoid tumour occurring in the nasal septum.

Carcinoid tumour was first described in 1888 by Lubarsch,[4] who found multiple tumours in the ileum during autopsies of two patients. It was not until 1907 that a German pathologist by the name of Oberndorfer described these lesions as carcinoid tumours.[5]

The annual incidence of carcinoid tumours is reported to be approximately two to three per 100 000. Due to the indolent nature of these tumours, the actual incidence could be higher. The majority occur between the ages of 60 to 65 years.[6]

Carcinoid tumours have been reported in a wide range of organs, but the majority occur in the gastrointestinal tract, followed by the bronchopulmonary complex.

Carcinoid tumours are a relatively uncommon subset of neuroendocrine tumours. They are described as neuroendocrine as they have the ability to produce endocrine products such as serotonin.

Carcinoid tumours arise from either the argentaffin cells of Kultchitzky or the amine and precursor uptake and decarboxylation cells. They predominantly occur in the small bowel, especially the appendix, rectum and terminal ileum. They are classified according to their embryonic origin (i.e. foregut, midgut or hindgut) and histological characteristics.[7] In histological terms, they are classified as well differentiated (typical) or moderately differentiated (atypical). Atypical tumours are more aggressive and behave like a malignant tumour, whereas typical tumours are often indolent and benign.[8]

Case report

An 83-year-old woman presented to the ENT out-patients department at Walsall Manor hospital with a six-month history of left-sided nasal blockage with anterior rhinorrhoea. She had no other significant past medical history and was a non-smoker.

Flexible nasoendoscopy revealed a polyp in the left nasal cavity, which was thought to be extending from the left middle meatus. Examination of the right nasal airway and post-nasal space was unremarkable, as was the remaining ENT examination.

A magnetic resonance imaging scan of the nose and paranasal sinuses was requested. This showed a soft tissue mass which appeared to erode the adjacent upper nasal septum and ethmoid cell walls (Figure 1). There was also some erosion of the left cribriform plate, but the adjacent cerebral gyrus was not compressed. The medial wall of the left orbit was intact. The remaining paranasal sinuses were normal.

In order to assess the exact degree of bony invasion of the tumour, a computed tomography (CT) scan was requested. This confirmed the presence of a soft tissue mass eroding the nasal septum and extending into the left anterior ethmoid cells (Figure 2). However, the CT scan showed that the cribriform plate was intact and not eroded by the tumour.

The patient underwent surgical removal of the polypoid mass, together with a wide upper septectomy.

From the ENT Department, Walsall Manor Hospital, UK.
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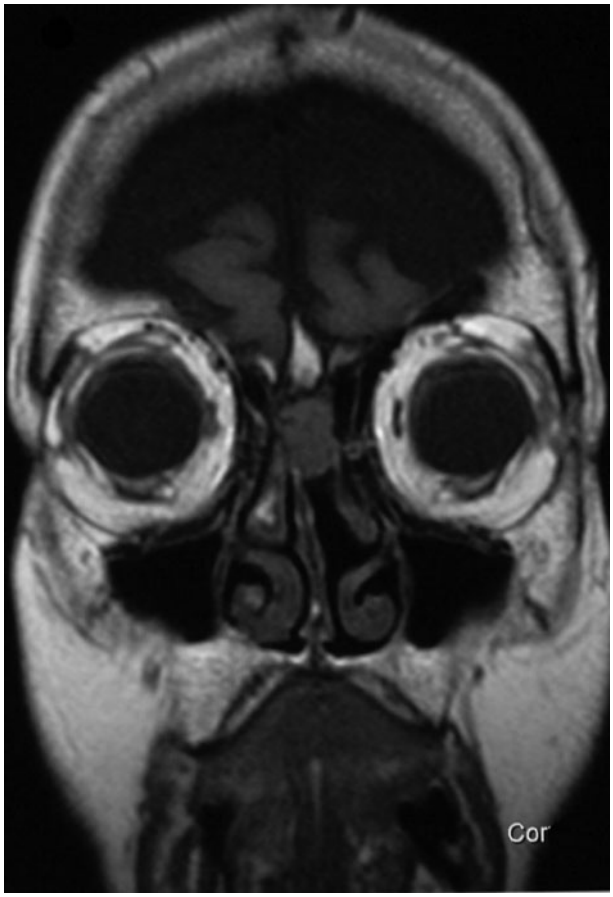

FIG. 1
MRI scan showing the soft tissue mass arising from the nasal septum.

The histological appearance of the operative specimen was in keeping with a carcinoid tumour, and the cells were positive for chromogranin A (a marker for neuroendocrine tissue) (Figure 3).

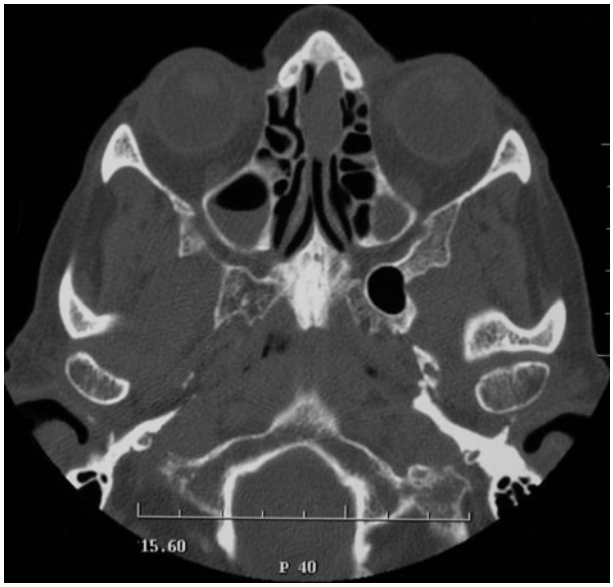

FIG. 2
CT scan showing no invasion of the cribiform plate.

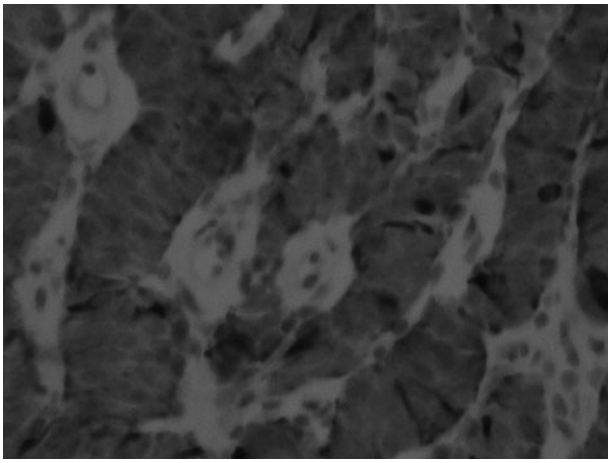

FIG. 3
Photomicrograph showing cells staining positive for chromogranin A (a marker for neuroendocrine tissue) (×20).

A 24-hour urinary 5-hydroxyindole acetic acid test was negative, and a chest X-ray did not show any other lesions or cardiomegaly.

Since surgery, the patient has shown no symptoms suggestive of carcinoid syndrome, such as flushing, bronchospasm or diarrhoea. Furthermore, she has displayed no signs or symptoms of liver involvement. At the time of writing, four years from initial presentation, she remained free of recurrence on nasoendoscopy (Figure 4).

Discussion

Tumours of the nasal septum are rare. There have been reported cases of squamous cell carcinoma, adenocarcinoma, lymphoma,[9] pleomorphic adenoma[10] and schwannoma[11] of the nasal septum. Carcinoid tumours arising in extragastrointestinal locations are also rare. To our

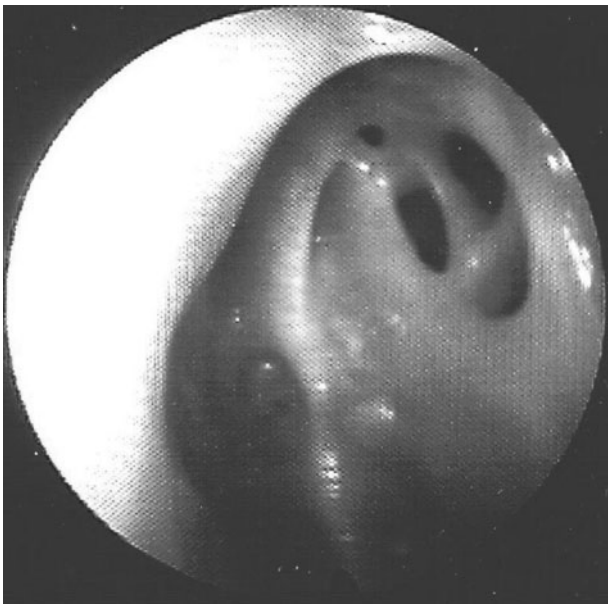

FIG. 4
Post-operative view looking through the right nasal airway with a flexible nasoendoscope, showing complete resection of the tumour.

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knowledge, there have been no previously reported cases of carcinoid tumours of the nasal septum.

Carcinoid tumours belong to the family of neuroendocrine tumours, which are usually slow-growing and have distinct histological and clinical characteristics. In histological terms, they are characterised by a positive reaction to silver stains and to markers of neuroendocrine tissue, such as chromogranin.[12] Typical carcinoid tumours contain small cells with regular, well rounded nuclei, and have five patterns of growth: insular, trabecular, glandular, undifferentiated and mixed.[13] In contrast, atypical tumours show increased nuclear atypia and greater mitotic activity.

Under the electron microscope, carcinoid tumours are typically found to contain membrane-bound neurosecretory granules. These granules contain the hormones and the biological amines that may be subsequently released by the tumour.

Carcinoid tumours commonly release serotonin, which is produced from the precursor 5-hydroxytryptophan. Five-hydroxyindole acetic acid, which can be used to monitor carcinoid tumours, is produced by metabolism of serotonin in the liver and is then excreted in the urine.[13] Carcinoid tumours also have the ability to secrete a variety of biologically active amines such as peptides (including serotonin), bradykinin, histamine, tachykinins and prostaglandins.

- **Carcinoid tumours commonly occur in the gastrointestinal tract and lungs**
- **Cases have been reported in the larynx, parotid and middle ear**
- **This is the first reported case of primary carcinoid tumour in the nasal septum**
- **Treatment of choice for carcinoid tumour in the nasal septum is primary surgery. Regular follow up is mandatory to monitor for metastases to the liver and resultant carcinoid syndrome**
- **Post-operatively, our patient remained free of local recurrence and distant metastases, four years from initial presentation**

Carcinoid tumours have often been diagnosed due to incidental findings at surgery or at autopsy. This is largely due to their slow-growing nature and non-specific presentation. Clinically, most carcinoid tumours remain asymptomatic until metastasis occurs.

Approximately 10 per cent of patients with carcinoid tumours present with carcinoid syndrome. This is characterised by the presence of flushing, bronchospasm, diarrhoea and right heart failure. When patients present with such symptoms, they are caused by metastases in the liver.[14] Even rarer is the development of carcinoid crisis. This occurs when the tumour outgrows its blood supply and large amounts of mediators are released into the blood stream, which is potentially life-threatening.[15]

Management of a patient presenting with any unilateral polyp in the nasal cavity should include an initial biopsy to exclude a malignant cause. As with other cases of carcinoid tumours encountered in the head and neck region, the choice of treatment in our case was surgical resection. Our decision (especially in view of the patient's age and lack of involvement of the cribriform plate) was to undertake wide surgical excision of the carcinoid tumour via an upper nasal septectomy, with regular follow up. Due to the anatomical site of the tumour, it was fairly easy to monitor signs of local recurrence with a flexible nasoendoscope. Our patient has not shown any signs or symptoms of liver metastases.

The management of patients with liver metastases includes chemoembolisation of these metastases. The use of radiofrequency ablation has been reported, resulting in decreased symptoms in all patients; however, the follow-up time in this study was only six months.[16]

Surgical resection of liver metastases may be of benefit in patients with limited hepatic disease. Such surgery has resulted in long-term relief of symptoms and prolonged survival in selected patients.[17–19]

Somatostatin analogues have been used not only for treatment but also for diagnosis of carcinoid tumours.[12] Octreotide is often used to relieve the symptoms of carcinoid syndrome.

The use of chemotherapy agents such as 5-fluorouracil, doxorubicin and streptozocin has not been successful. However, chemotherapy can be used in combination with surgery, chemoembolisation and somatostatin analogues.[20]

Carcinoid tumours are not particularly radiosensitive; thus, radiation does not usually play a therapeutic role except, rarely, for the treatment of bone metastases.[21]

It is difficult to predict the long-term prognosis of a primary carcinoid tumour in the nasal septum. However, our patient remained free of tumour four years after initial presentation.

Conclusion

To our knowledge, and following a thorough literature review, our case represents the first report of primary carcinoid tumour of the nasal septum. Our management comprised wide surgical excision of the tumour and then regular follow up. It is important that the patient is closely observed for signs and symptoms of metastatic disease.

The possibility of primary carcinoid tumour of the nasal septum should be considered by the surgeon when encountering a mass within the nasal septum, because timely recognition and intervention are critical for successful treatment.

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Address for correspondence:
Ms T Galm,
Walsall Manor Hospital,
Moat Road,
Walsall WS2 3PS, UK.

E-mail: tiragalm@yahoo.com

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