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Pathology in Focus

Teratocarcinosarcoma of the nose, paranasal sinuses and nasopharynx

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Abstract
Teratomas affecting the head and neck are rare tumours of ambiguous origin. The benign form is seen in children and mostly affects the nasopharynx. Malignant tumours in the upper jaw have been reported rarely. They are almost exclusively seen in the adult male. They are highly aggressive tumours that are difficult to diagnose pre-operatively due to the varied histological patterns. Treatment options available are surgery, radiotherapy or a combination of both. Despite intensive treatment, the prognosis is still poor. A rare case of teratocarcinosarcoma of the nose, paranasal sinuses and nasopharynx in a 25-year-old male is presented. The tumour had a non-germ cell tumour origin. The clinical features, pathologic characteristics and treatment are detailed. It was managed by surgery, radiotherapy and chemotherapy.

Key words: Teratocarcinosarcoma; Nose; Paranasal Sinuses; Nasopharynx

Introduction

Of the malignant tumours that can affect the nose, paranasal sinuses and nasopharynx, teratomas are extremely rare. They constitute five per cent of all germ cell tumours both benign and malignant.[1]

Malignant teratomas arising outside of the gonads are rare events[2] and their clinical presentation, histiogenesis and treatment has been an enigma to the treating doctor. Only a handful of similar cases has been reported in the past from the upper jaw.

Case report

A 25-year-old male, presented to the out-patient department with complaints of left-sided nasal obstruction for four months, and bulging of the left eye and diminution of vision for three months. He also had had several episodes of bleeding from the nose and a frontal headache. Nasal obstruction had been insidious in onset, initially only in the left but later bilateral. Fifteen days prior to admission the patient noticed that a mass had filled his nose and protruded outside it. He had also noticed a mass in the oropharynx, that was causing difficulty in swallowing.

Examination showed a red, friable mass projecting for about 3 cm from the left naris (Figure 1). The mass was seen to fill the entire left nostril. Medially it had pushed the septum completely to the opposite side. Posteriorly it had filled the nasopharynx and displaced the soft palate to present in the oropharynx, just reaching the upper limit of the hypopharynx. Inferiorly it had eroded the hard palate to present in the oral cavity. There was destruction of the lateral wall of the nose with obliteration of the nasolabial fold on the left. There was fullness over the left maxilla. The left eye showed proptosis, with the eyeball displaced laterally and inferiorly. The eye movements were restricted in all directions. Pupillary reaction was present but vision was restricted to perception of light only. The fundus showed primary optic atrophy. Examination of the rest of the cranial nerves and neurological system was normal. No neck nodes were palpable.

A routine haemogram showed a haemoglobin value of 5.1 g/dl, which was then corrected by pre-operative blood transfusion and haematinics. A computed tomography (CT) scan of the nose and paranasal sinuses showed a nonhomogeneous, enhancing mass occupying the entire

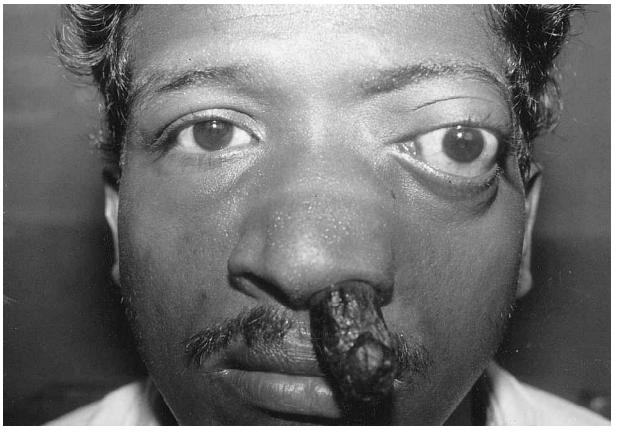

Fig. 1
Pre-operative photograph of the patient showing tumour protruding from the left naris.

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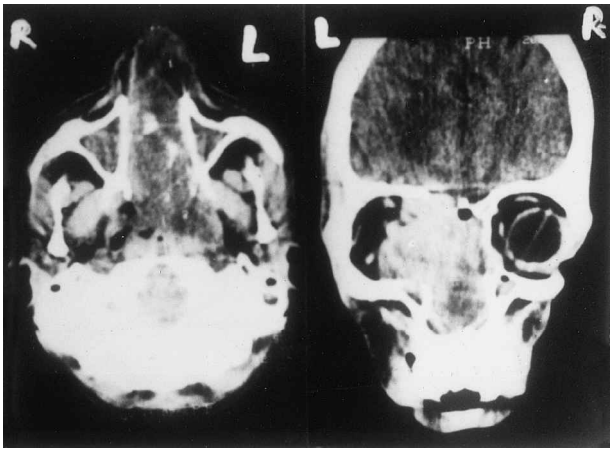

FIG. 2

Left half – Axial CT scan showing tumour in the left nose extending to ethmoids, sphenoid and nasopharynx. Right half – Coronal CT scan of the nose showing tumour in the left maxilla, ethmoids, and bony defect in the cribriform plate without intracranial extension.

left nostril and nasopharynx (Figure 2). The tumour had eroded the ethmoids, cribriform plate, the medial wall of the left maxilla, the anterior and lateral walls of the sphenoid and the hard palate. No evidence of intracranial extension was seen. Based on clinical features and radiological findings, a diagnosis of malignancy of the nose, paranasal sinuses and nasopharynx was made.

Two consecutive biopsies failed to give a conclusive report, as they consisted of only extensive necrotic material with no viable cells. During this period it was noticed that the tumour was growing aggressively, with a rapid increase in size in all directions and increasing proptosis. The patient lost vision progressively becoming blind in the left eye. The protruding necrotic mass from the nose and oropharynx fell off. A third biopsy also showed extensive necrosis but in some areas the tumour was lined with vascular channels and the pathologist considered a differential diagnosis of a variant of an angiofibroma or bacillary angiomatosis. These being benign lesions, it was decided to operate.

In the meantime, the patient developed malaria for which he was administered antimalarial drugs – chloroquine for three days and primaquine for 15 days. Three days after the start of treatment, the patient developed massive bleeding from the lesion, which he aspirated. Bleeding and aspiration was so severe that it threatened the life of the patient. An emergency bedside tracheostomy was performed and the patient revived. Antimalarial therapy was continued. One week after completion of antimalarial therapy, a transfemoral external carotid angiogram was done. This showed a midline nasal mass supplied by the facial and internal maxillary artery on the left and by the lingual and facial artery on the right. Embolization of the same was unsuccessful.

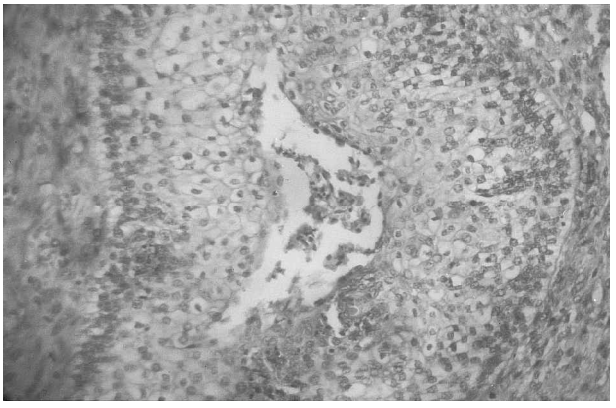

FIG. 3

Clear-cell squamous epithelium with a thin basal zone resembling fetal oral mucosa (H&E; ×200)

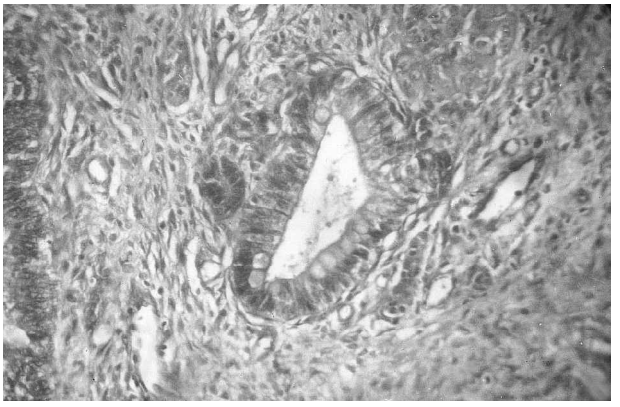

FIG. 4

Gland lined by mucin containing tall columnar cells and goblet cells resembling gut mucosa (H&E; ×200)

The next day, the patient was operated on. The neurosurgical team was available for intervention in case of an intracranial extension of the tumour. Initially it was prepared for external carotid artery ligation in the event of uncontrollable bleeding during dissection. The left carotid was exposed and stay sutures were placed around it without ligating it. The tumour was approached through a Weber Ferguson incision with subciliary extension. The mass was found to extend from the nose into the maxilla, ethmoids, sphenoid, pterygopalatine fossa and overhang into the nasopharynx, conforming with the CT scan findings. The entire tumour mobilized easily. It was highly friable and was removed with minimal bleeding. Considering the pre-operative diagnosis of a benign tumour and as the orbit was not involved, it was only decompressed. The surgical defect was closed by an obturator and the cavity was packed.

The specimen was sent for histopathological examination. Grossly the tumour was irregular and friable. A cut section of the tumour showed predominantly dark brown haemorrhagic areas and partly pale brown to whitish viable tumour. Microscopic examination showed features of a malignant teratoma with both epithelial and connective tissue elements. The epithelial components consisted of well-defined nests of squamous epithelium with clear cytoplasm, with some nests showing central keratinization (Figure 3). The connective tissue element had large glandular structures with tall columnar cells, clear cytoplasm, cilia and uniformly rounded nuclei (Figure 4). Some glands were lined by oncocytic epithelium and others with primitive gastrointestinal epithelium. Distinct neuroepithelial elements were also seen with tubercules and rosettes in a neurofibrillary matrix (Figure 5). The mesenchymal component consisted of primitive spindle cells in a myxoid stroma. Focally the stroma was highly cellular with pleomorphism, mitosis, areas of infarction, haemorrhage and osteoid production in some areas – osteogenic sarcoma (Figure 6). Tumour markers were estimated for germ cell tumour origin. Beta human chorionic gonadotropin (HCG) was 1.0 mg/100 ml (normal less than 10 for a non-pregnant person) and α-feto protein was 4.5 mg/100 ml (normal 0–10).

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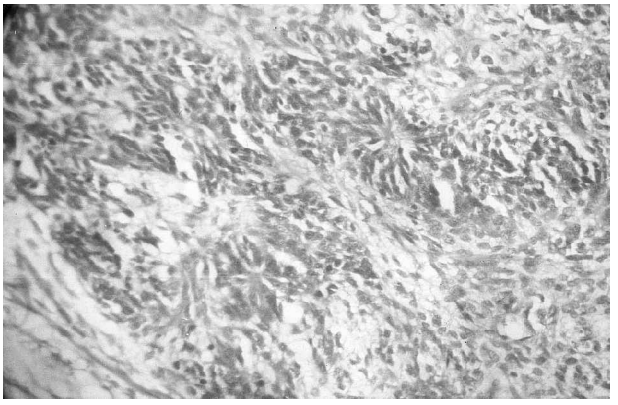

FIG. 5

Masses of neuroepithelial tissue with Homer–Wright rosettes containing central pink fibrillary processes (H&E; ×200)

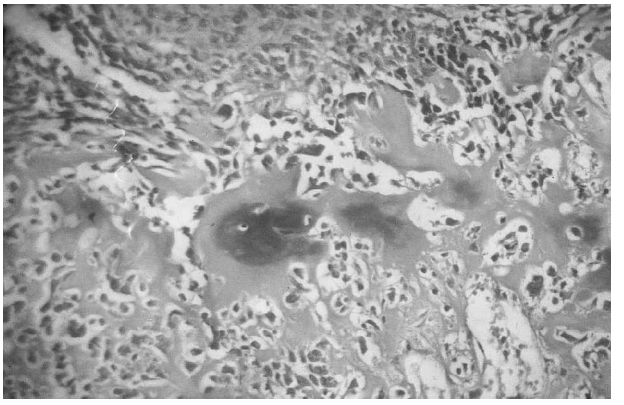

FIG. 6

Hyperchromatic, pleomorphic spindly sarcoma cells producing osteoid – osteogenic sarcoma (H&E; ×200)

Two days post-operatively the patient developed right-sided hemiplegia. CT showed a haemorrhagic infarct in the left temporal region. A neurology opinion was sought and they opined that the cause of the hemiplegia was embolization or the effect of the antimalarial drugs. Hemiplegia was managed conservatively. The wounds healed well and after 15 days the patient was referred for radiotherapy. 4500 cGy of teleradiotherapy was given in 15 hypofractionated doses. This was combined with cisplatin – 100 mg/m^2 for five days and etoposide – 600 mg for three days. The patient has been followed up for the last 13 months and he is symptom free for the nasal disease. There has been improvement in his hemiplegia, and he is able to move with support.

Discussion

Malignant teratoma was first documented by Patchefsky *et al.*[2] from the ethmoid sinus. Despite surgical treatment his patient survived for only three months.

Incidence

Teratomas of the nose and paranasal sinuses are rare events. Their benign counterparts are commonly seen in the nasopharynxes of children but rarely in adults.[2,3] They are the most common extragonadal germ cell tumours in childhood.[1] The malignant form is seen exclusively in adults with a clear male predilection.[4] Sites of occurrence in the head and neck include the soft tissues of the neck, sinonasal region, nasopharynx, oral cavity and orbit.[1] Previous terminologies given for this tumour include malignant teratoma, carcinosarcoma and malignant mixed tumour.

Clinical features

The clinical symptoms most commonly manifested are innocuous, usually nasal obstruction and epistaxis. The malignant nature of the lesion is inferable from its rapid growth.[5] In our case, there were two peculiarities:

(1) The massive haemorrhage from the tumour pre-operatively suggested a diagnosis of angiofibroma. A similar presentation for a teratocarcinoma has not been reported in the literature.
(2) The tumour itself on observation on a day to day basis was fast growing. A part of the mass protruding from the nose and oropharynx had become necrotic and fallen off. Such a presentation also has not been described previously.

Diagnosis

Generally the anatomical site of origin is impossible to discern due to its size and rapid growth. When the tumour is located high in the nasal cavity it may be mistaken for an aesthesioneuroblastoma.[2] All these cases have to be thoroughly investigated; CT scan being the investigation of choice for confirming the extent of disease. Pre-operative diagnosis by biopsy has often led to erroneous diagnoses such as craniopharyngioma, adamantinoma or inverted papilloma.[2,3] This is due to the varied histological pattern. Unless definite teratomatous areas are biopsied, the picture seen is likely to confuse the pathologist. In our case we faced a diagnostic problem as the first two biopsies were negative and the third biopsy was reported as a vascular lesion. It was only after the tumour was removed in toto, that a final diagnosis of malignant teratoma of the sinonasal region was made.

Cell of origin

The exact histiogenesis of the tumour is still in doubt. Possible cells of origin are the gonadal germ cell and neural crest.[1] In our case due to the absence of germ cell tumour areas on histopathological examination and normal serum values of germ cell tumour markers, we feel the tumour had a non-germ cell origin which is rare.

Histopathology

Histologically, the tumours are characterized by a mixture of epithelial and mesenchymal components including cellular elements with immature or embryonal characteristics. These tumours, consisting of an epithelial element and one or more mesenchymal components are variously termed teratocarcinosarcoma, carcinosarcoma, malignant teratoma, spindle-cell carcinoma, and pseudosarcomatous squamous cell carcinoma.[6]

Treatment

Treatment approaches recommended are a combination of aggressive surgery and radiotherapy.[1,4,5] It constitutes a challenge as these tumours are highly malignant and the majority of patients die in a short time. Abt *et al.* found radiotherapy to be of little benefit.[3] Subsequent observations especially the large series by Heffner *et al.* suggest that radiotherapy still has a role, although it cannot be relied on as a sole modality of treatment.[4] The role of chemotherapy for these tumours is still experimental. With aggressive treatment there is a chance for increased survival, with maximal survival described in literature being a three-year survival of 40 per cent.[4] We treated our

case by a combination of radical surgery, radiotherapy and chemotherapy and the patient is doing well even 13 months post-operatively.

Conclusion

Teratocarcinosarcoma will be encountered by the otorhinolaryngologist rarely. Pre-operative diagnosis is rarely possible by biopsy, as the histological pattern is varied and misleading. On a clinical suspicion of malignancy, aggressive surgical therapy has to be undertaken for increasing chances of survival. In diagnosed cases, post-operative radiotherapy and chemotherapy constitute optimal treatment.

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Dr C. Prasad takes responsibility for the integrity of the content of the paper.
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