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Malignant schwannoma of the nasal cavity and paranasal sinuses in a Nigerian

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Summary

An unusual case of malignant schwannoma with involvement of the forehead, external nose, right nasal cavity, paranasal sinus system (bilateral frontal sinus, right ethmoidal sinus), right orbit and anterior cranial fossa is reported in a Nigerian. Malignant schwannomas of the paranasal sinus are extremely rare, as only 20 well-documented cases have been previously published in English literature. No report in black Africans has been found in extant literature. The clinical features of this tumour are presented with detailed management. The patient had a wide surgical resection of the lesion with reconstruction of the resultant fronto-nasal defect using forehead musculofascial flap plus full thickness skin graft and adjuvant radiotherapy with satisfactory outcome. The good result of combined surgery and radiation regimens in this case demonstrates the usefulness of adjuvant radiation therapy in this condition.

Keywords: *Schwannoma, malignant, nasal, sinuses.*

Résumé

Dans un cas irrégulier du schwannien bénigne associé au front , nez , cavité nasale droite, au système sinus paranasal (sinus bilatéral frontal,le sinus droit ethmoïdal), l'orbite droite, et la fosse craniéne antérieure est rapporte au Nigeria.Les schwanniens bénignes du sinus panasal sont extrêmement rare, seulement 20 cas documentés dans la littérature anglaise sans aucun rapport sur les noirs africains. Les caractéristiques cliniques de cette tumeur sont présentées avec les soins détaillés. Le patient avait une résection chirurgicale large de la lésion avec une reconstruction du defect frontonasal utilisant du flap musculo-faciale plus la graphe de la peau épaisse et une radiothérapie entraînant un résultat satisfaisant. Le bon résultat de la chirurgie combinée et les régiments de radiation démontrent l'utilité d'adjuvant de radiation thérapeutique dans cette condition.

Introduction

Malignant schwannomas, also known as malignant peripheral nerve sheath tumours arise from somatic and autonomic components of cranial and spinal nerves and account for 5-10% of all sarcomas [1]. The lower extremity is the most commonly involved site. However as much as 25-40% of schwannomas (benign and malignant) occur in the head and neck region [2,3]. Malignant schwannomas may arise sporadically or in association with von-Recklinghausen's disease or neurofibromatosis type 1(NF-1).

The sporadic form most commonly arises in persons aged 40-60 years and females are affected more than males whereas tumours arising in association with NF-1 most commonly occur in those aged 20-40 years and males are affected more than females [2]

Two percent of individuals with NF-1 are at risk of developing malignant schwannomas. A painful progressively enlarging tumour usually heralds the onset of malignant transformation in these patients [4]. Most sporadic examples of malignant schwannomas are idiopathic in nature, although a few examples have followed radiotherapy [5].

Relatively little is known of the molecular genetic alterations that underlie the genesis of malignant schwannomas. DNA sequencing studies have revealed mutations in P53, while an immunohistochemical study of these neoplasms revealed over expression of the p53 gene product [6]. Immunopositivity for this gene product was associated with a shorter median patient survival, which suggests that P53 gene mutation is required for progression of neurofibroma to malignant schwannomas [6]. Other cytogenetic studies have elucidated specific chromosomal aberrations including t(X;12)(q22;q24), t(2;4)(q35;q31) [7].

We present a case of mid-facial malignant schwannoma involving the right nasal and paranasal sinuses in a Nigerian.

Case report

The patient was a 31-year-old Nigerian female who presented with a 16-year history of a progressive mid-facial swelling, which commenced at the right lower eyelid and extended towards the medial canthus across the nasal root to involve the frontal region.

The swelling was painless, initially small and continued to increase in size to attain its large size after presentation. It displaced the right globe with a gradual loss of vision and complete blindness of the right eye about 2 years prior to presentation. There was no history of seizure, or personality changes. There was no bleeding or

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ulcer associated with the tumour. There were no other symptoms referable to the ear, nose, and throat and there were no associated systemic symptoms.

The patient's vital signs were normal; with a pulse rate of 84 beats per minute, blood pressure of 120/80 mmHg and respiratory rate of 24 per minute. Ear, nose and throat examination revealed no abnormality. However there was a huge, firm lobulated mass measuring 20 cm (transverse) by 18 cm (vertical) involving the frontal region, right orbit, nasal root and bridge crossing towards the left orbit (figures 1a, b,).

Fig. 1a: Pre-operative appearance (front view)

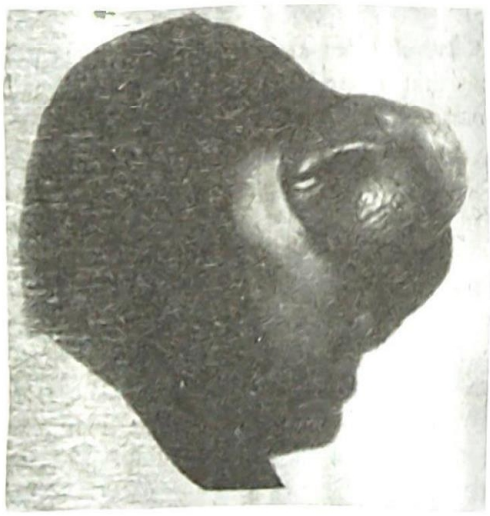

Fig. 1b: Pre-operative appearance (right lateral view)

Her mental status and long tracts were found normal. Apart from anosmia and right-sided blindness, peripheral nerve examination was normal. Other organ systems were essentially normal on clinical examination.

The computerised axial tomographic (CT) scan of the paranasal sinuses and brain revealed a huge lobulated soft tissue mass over the right facial region (figure 2a). The axial and coronal cuts showed a lobulated soft tissue mass of mixed density, with peripheral enhancing tissue nodules and central hypo-density. The mass had extended into the right nasal cavity and the anterior ethmoidal sinus. There was associated deviation of nasal septum to the left and erosion of the margins of the right nasal cavity as well as the ethmoid and medial walls of the right orbit (figure 2b). The right globe was displaced laterally with retrobulbar compression, but apparently not involved by the tumour. The floor of the anterior cranial fossa in the mid-line and on the right showed erosion with slight intracranial extension of the mass involving the frontal lobes. The remaining brain tissues and ventricles appeared normal. Other investigations including haematological tests and chest radiographs were essentially normal. Pre-operative multi-disciplinary sessions were held to discuss the management of this patient.

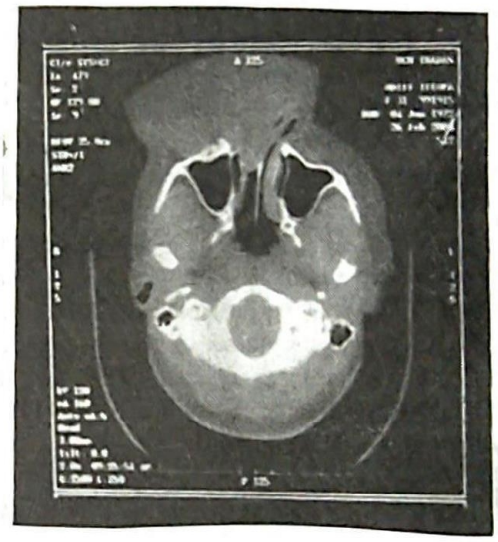

Fig. 2a: CT scan of the sinuses and brain (axial view)

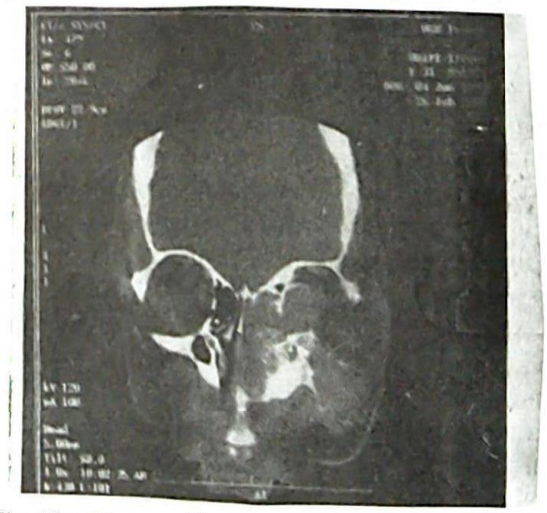

Fig. 2b: CT scan of the sinuses and brain (coronal view)

The patient had a 1.5 cm skin margin total excision of the mid-facial tumour, with bilateral frontal sinus, right ethmoidal and right nasal clearance under hypotensive general anaesthesia. A reconstruction using a turn over left forehead musculofascial flap plus full thickness skin graft of the right fronto-nasal defect was done (figure 5). The right globe was left in place for cosmetic purpose, although there was no vision.

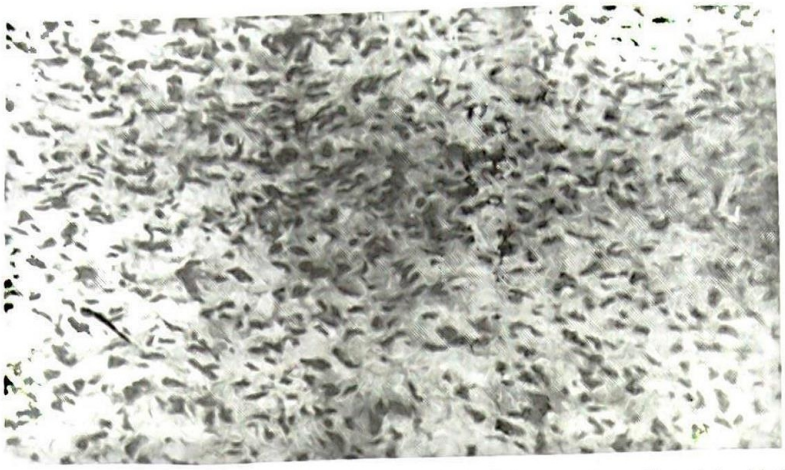

Fig. 3: Photomicrographs of microscopic tissue specimen showing pleomorphic cells with buckled nuclei

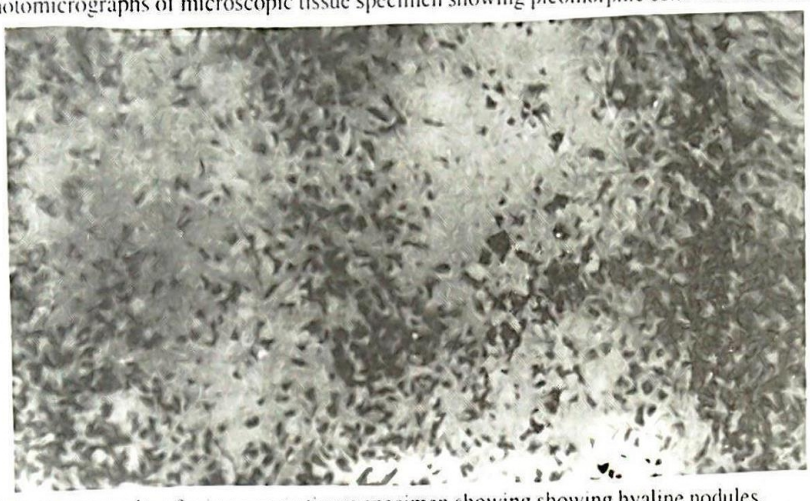

Fig. 4: Photomicrographs of microscopic tissue specimen showing showing hyaline nodules.

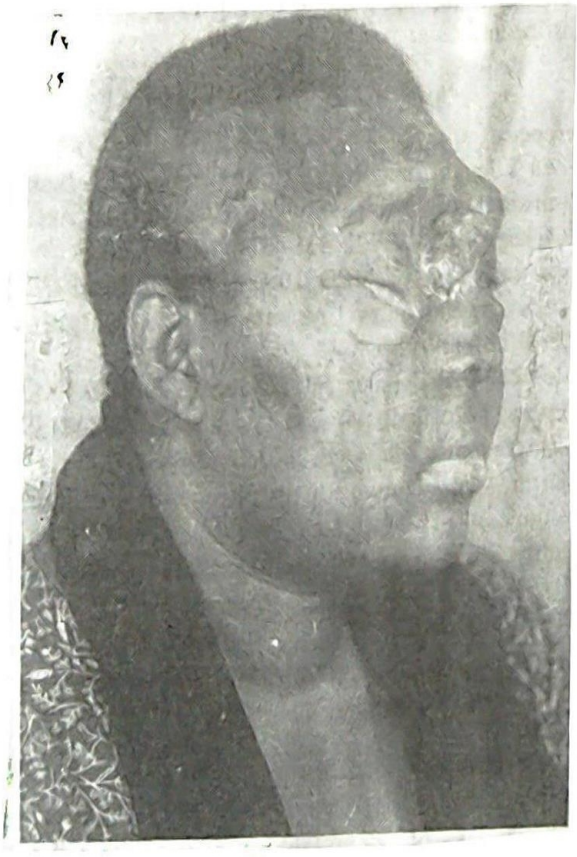

Fig. 5: Post-operative appearance (front view)

The tumour removed measured 16cm by 12cm by 11cm in size and weighed approximately 800 grams. The cut sections revealed a well circumscribed encapsulated multilobular greyish orange appearance with focal areas of haemorrhage. Gritty sections were felt in some areas. Histology showed a poorly circumscribed fasciculated spindle cell neoplasm composed of moderately pleomorphic cells having buckled nuclei (fig. 3) and pale eosinophilic cytoplasm with focal hyaline nodules (fig.4). A diagnosis of malignant schwannoma was made.

Patient subsequently had post-operative radiotherapy of 50 Gy in 25 fractions over 5 weeks and remains well with no sequelae 2 years after surgery.

Discussion

Malignant schwannoma is an aggressive neoplasm with a broad spectrum of histological patterns and despite advances in diagnostic techniques, the natural history of this tumour remains uncertain [8].

The most common site of origin of malignant schwannomas in the head and neck region is the neck followed by the nasal cavity and paranasal sinuses, nasopharynx, oral cavity, orbit, cranial nerves and larynx. Malignant schwannomas usually present insidiously and thus are often diagnosed incorrectly or after lengthy delays.

Clinical symptoms vary according to the site involved, but the most common presenting symptom is a painful enlarging mass [2].

In this case, the malignant schwannoma presented as a painless enlarging mass over a 16 year period involving the soft tissue of the forehead, nasal root and bridge, both the frontal sinuses, right ethmoidal sinus, and right nasal cavity. Invasion of the anterior cranial fossa and right orbit resulted in total visual loss, thus confirming the aggressive nature of this tumour.

This appears to be the first report documented in the literature of malignant schwannoma involving the head and neck in a black African. The age and sex of this reported case of malignant schwannoma are in consonance with a sporadic neoplasm, since sporadic tumours are commonly seen among females and middle age patients, and in view of the absence of a family history suggestive of type 1 neurofibromatosis (NF1).

As many as 50% of patients with malignant schwannomas have evidence of NF-1 or a positive family history and about 5-15% of patient with NF-1 have malignant schwannoma [2,9]. Therefore, a diagnosis of malignant schwannoma should suggest the possibility of NF-1, and a rapidly enlarging painful mass in a patient with NF-1 would suggest a malignant schwannoma.

A correct diagnosis with a CT Scan to delineate the tumour extent is imperative. In addition, angiography has been found useful in its diagnosis in which schwannomas present a definitive vascular pattern. In patients with head and neck tumours whose angiographic findings include a pattern of moderate hypervascularity, tortuous tumour vessels and scattered contrast puddles without arteriovenous shunting or vascular encasement; schwannomas should be suspected [10,11].

Microscopically, malignant schwannoma cells appear spindled, contain scanty cytoplasm, and are oriented in sweeping fascicles that imitate a herring bone pattern with cellular pleomorphism, elongated, wavy and buckled nuclei. Nuclear palisading may be present and characteristically may display hyaline bands and nodules as were found in this case. Immuno-histochemical studies for nerve sheath differentiation reveal positivity for S-100 protein, leu-7 and myelin basic protein. S-100 immuno-reactivity is focal and scattered in 50-90% of malignant schwannomas. The other two antigens show immunoreactivity in approximately half of the tumours [13].

Treatment of malignant schwannomas is primarily surgical with wide excision where possible and adjuvant radiation therapy is often used because most of these tumours are high grade [2,3,8,9,12]. Excessive bleeding is always associated with this tumours during surgery hence embolization during angiography is a useful and safe presurgical adjunct in the treatment of vascular schwannomas [10]

Pre-surgically, in this patient, embolization could not be carried out; hence a hypotensive anaesthetic technique was performed to maintain the patient's mean arterial blood pressure at between 60-65 mmHg during the course of surgery in order to minimize excessive blood loss. The total blood loss was about 1 litre at the end of the three and half hours surgery.

The modalities of the transfusion support including combined transfusion therapy with an autologous unit (preceding erythropoietin therapy) and homologous unit is being addressed in another paper.

Adjunct radiation therapy consisting of 50 Gy in 25 fractions was given. Chemotherapy may have a role in the treatment of an in-operable disease, recurrent disease or disease that persists despite initial therapy.

The outcome appears to differ with the clinical setting in which the tumours arise. In sporadic malignant schwannomas, reported 5-year survival rates are 50-75% whereas survival rates for malignant schwannomas associated with NF-1 are 15-30% [2,8]. This case is still being followed up. However the present post-operative status shows that the tumour is under control.

In conclusion we have presented a case of malignant schwannomas with involvement of the forehead, external nose, nasal cavity, paranasal sinus system, orbit and anterior cranial fossa which was successfully treated surgically and with adjunct radiotherapy. This is a rare surgical problem and it is our view that in this case the prognosis is good since the tumour is of the sporadic type without preceding neurofibromatosis.

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