Maxillary haemangiopericytoma: A case report

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Summary

Background: **Haemangiopericytoma is a very rare slow-growing vascular tumour with a variable malignant potential, constituting less than 1% of all neoplasms. It may arise from any blood vessel and in any organ of the body. Primary haemangiopericytoma of bone is even rarer, constituting about 0.1% of bone tumours. The tumour is extremely rare in Africans and particularly in the head and neck region.**

Study design: **We describe the case of a 66-year old Nigerian with haemangiopericytoma of the maxilla, who presented with a recurrent but painless jaw mass.**

Results: **Surgical resection of this tumour is potentially bedevilled with the risk of torrential haemorrhage and high rate of recurrence. This risk may be substantially reduced by wide surgical resection with a careful microscopical examination of the resection margins and the institution of adjuvant radiotherapy in incompletely resected tumours. Chemotherapy has no known role in the management of haemangiopericytoma. Postoperative radiation therapy appears to be effective against tumour recurrence.**

Conclusion: **Even then, long-term follow-up is essential in all cases. To our knowledge, this is the first report of this entity in an African.**

Keywords: ***Haemangiopericytoma, Embolisation, Afferent vessel ligation, Radiotherapy Chemotherapy, Prognostic factors.***

Résumé

Introduction: L'hémangiopericytome est une tumeur vasculaire très rare qui se développe lentement avec un variable malin potentiel constitute moins de 1% de tous néoplasmes. Elle peut se présenter à partir de n'importe quel vaisseau du sang et dans n'importe quel organe du corps. L'hémangiopericytome primaire d'os est également plus rare, constitute 0,1% des tumeurs d'os. La tumeur est extrêmement rare chez les africains en particulier dans la region de la tête et du cou.

Plan d'étude: Nous présentons le cas d'un Nigerian age de 66 ans atteint dd'hémangiopericytome du maxilla, qui s'est présenté atteint de la masse machoire sans douleur mais récurrente.

Résultat: Résection chirurgicale de cette tumeur est potentiellement en proie à risqué d'une hémorragie torrentielle et un taux élevé de récurrence. Ce risque pourrait être en baisse considerablement par une résection chirurgicale très large avec un examen microscopique de terminaux d'une résection soigneux et l'institution de la radiotherapie adjuvante dans des tumeurs qui ne sont pas complète dont on n'a pas fait des résection chirurgicales complètes. La chimiotherapie n'a pas un role connu dans la prise en charge d'hémangiopericytome. La thérapie des radiations postopératoire parait être efficace contre la récurrence de tumeur.

Conclusion: Soins post-hospitaliers à long terme est necesssaire dans tous les cas. D'après nous , ceci est le premier rapport de cette entité chez un africain.

Introduction

First described by Stout and Murray in 1942[1], haemangiopericytomas are rare vascular tumours thought to originate from the pericytes of Zimmermann, contractile cells normally found around capillaries [2]. Haemangiopericytoma can occur in both primary and secondary forms, the primary lesions may be intramedullary or periosteal [3]. Theoretically, owing to the fact that blood vessels, from which they originate, are ubiquitous, haemangiopericytomas can occur in any part of the body, but they occur most commonly in the pelvis and the lower extremities [2]. Overall they represent less than 1% neoplasms arising from all organs and tissues of the body. Primary bone haemangiopericytoma is rare, constituting about 0.1% of Primary malignant bone tumours [2]. The occurrence of the tumour is rarer still in the bones of the

Fig. 1 *Frontal view of the patient at presentation.*

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head and neck region [4]. For example, only 52 cases of haemangiopericytoma had been documented in the literature between 1949 and 1990 out of which only two originated from the maxillary sinus [5]. Similarly, up till 1995, only 10 cases of primary haemangiopericytoma involving the jaw bones had been reported in literature [6]. The most common sites in the head and neck are the nasal cavity, and the paranasal sinuses, and unusually, the orbital region, the parotid gland and the neck[3,5]. We have not encountered any report of this tumour occurring in the bones of the head and neck region in an African. Hence we describe a case of haemangiopericytoma of the maxilla seen by us in a Nigerian woman.

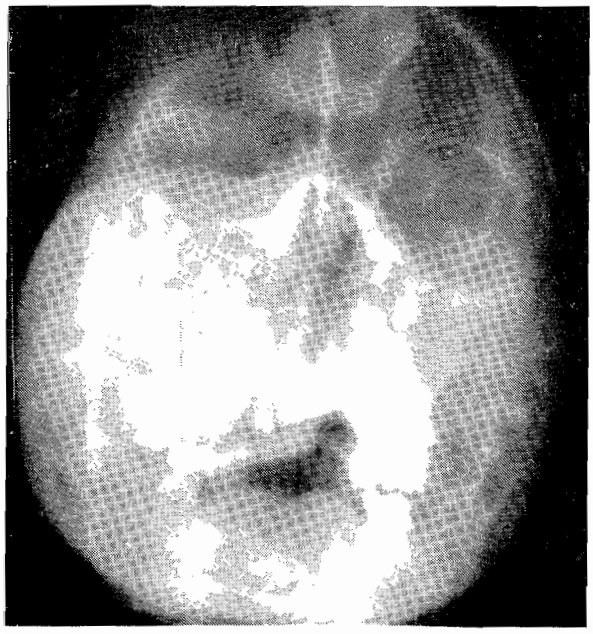

Fig.2 *Occipitomental view of the skull taken Note the radio-opaque right maxillary antrum.*

Case report

A 66-year old Nigerian female presented herself for treatment at the Maxillofacial Unit of the Obafemi Awolowo University Teaching Hospitals Complex, Ile-Ife, Nigeria, on March 2, 1999 with a principal complaint of recurrent painless swelling on the right side of her face (Fig.1). She first noticed the swelling about 9 years previously when she experienced a toothache on that side of her face. A mobile tooth was subsequently extracted leaving behind an unresolved swelling. Five months later, she consulted an oral surgeon in another hospital and was operated upon to remove the mass. The excised mass was not subjected to pathological examination. However, the swelling recurred about five years after the first operation. She sought help in our centre two years after the recurrence. She reported a subjective gradual weight loss, as the swelling grew bigger. She had never smoked but she drank alcohol (beer) occasionally. She habitually chewed ground tobacco (snuff) for an unspecified number of years but this habit was discontinued after she had the first operation.

The patient, though ill looking was not febrile to touch and all other vital signs were essentially within normal limits. Examination of the head and neck region showed a tense shiny skin overlying a firm, warm, lobulated and non-pulsatile swelling. There was obliteration of the right nasal fold. Intra-orally, the swelling extended from the upper left lateral incisor to the right maxillary tuberosity region with bucco-palatal expansion. Aspiration of the swelling yielded a bloody fluid, which did not clot even though the swelling did not transilluminate.

There was impaired breathing and there was a blood-stained purulent discharge intra-orally. The submandibular lymph nodes were bilaterally enlarged, freely mobile and firm in consistency. There was no paraesthesia over the region of the distribution of the right infra-orbital nerve. The oral hygiene was fair and there were five mobile teeth (upper right canine, upper right and left lateral and central incisors). Neurologic examination and other systems revealed no abnormalities.

Radiographs of the jaws disclosed a large soft tissue mass in the right maxillary region (Fig.2). The underlying maxillary antrum was opaque with poorly defined walls thereby giving the appearance of a widened antrum.

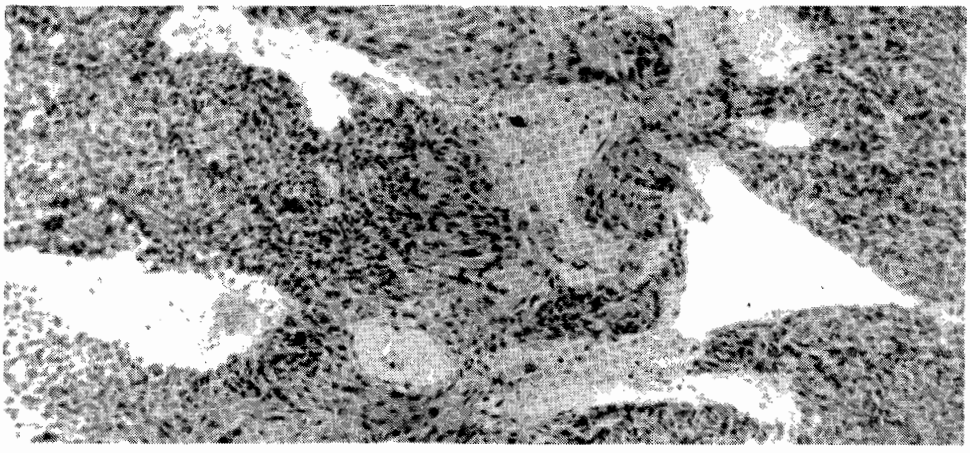

Fig.3a *Showing a highly vascular tumour with large sinusoid-like spaces which are surrounded by pericytic cells.*

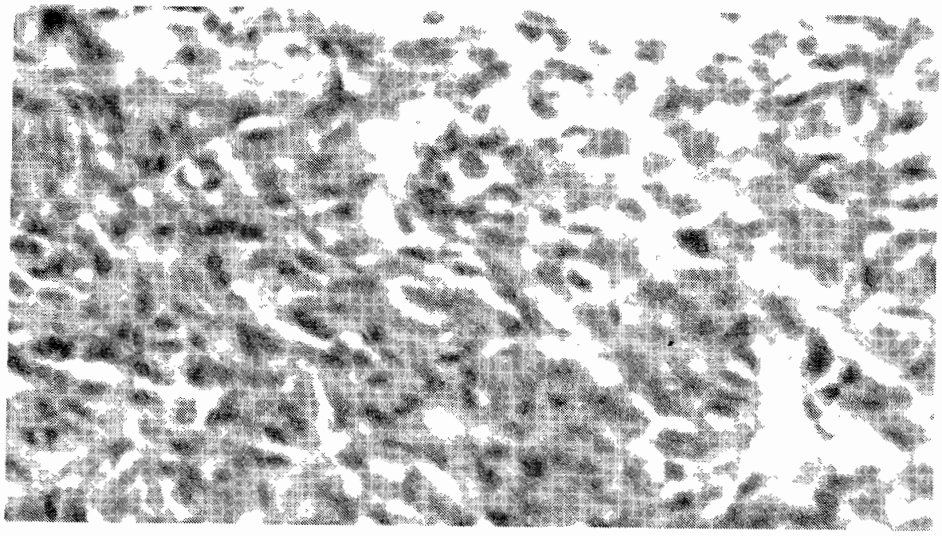

Fig. 3b *Showing pericytes around the abnormal vascular channels. Some of the pericytes show mitoses.*

There was suggestion of protrusion in the right half of the buccal cavity as evidenced by dense bulge of soft tissue with absence of the right half of the upper dental alveolar bone. There was partial occlusion of the right nasal cavity. The left maxillary antrum was hazy but the walls were not destroyed. Chest radiographs were within normal limits. Routine haematological and biochemical tests showed normal values. However, profuse bleeding followed an incisional biopsy that was done under local anaesthesia.

Histopathologic examination of the lesion showed a highly vascular tumour with large sinusoid-like spaces as well as capillaries about which were distributed plump and well-shaped pericytes. Some of the cells had clear cytoplasms while others had abundant eosinophilic cytoplasm. In certain areas, an eosinophilic matrix was being laid down. Some of the vascular spaces contained red blood cells. There were also extravasated red cells as well as focal deposits of haemosiderin, suggestive of old haemorrhages. Mitoses were frequent although most of them were normal Fig.3. The appearances were consistent with those of a malignant haemangiopericytoma.

She was scheduled for maxillectomy (radical surgical resection) to be supplemented with adjuvant radiotherapy. However, these were made difficult by the patient's difficult social circumstances. She attended only a few review sessions before she was lost to follow-up. However, we received reliable information that she passed –on in late March 2000. Autopsy was not carried out before burial.

Discussion

Pericytes are small, elongated spindle shaped pericapillary cells. They are located external to the endothelial cells and are separated by a basement membrane. For this reason, haemangiopericytoma may arise from any part of the body.

Haemangiopericytoma is a relatively slow growing tumour with locally invasive tendencies, variable malignant potential and a high rate of recurrence following conservative management [7]. All osseous haemangiopericytoma are now regarded as malignant but with varying metastatic potential[6] because metastasis has recently been described in some congenital types that were initially thought to be benign [8]. Although rare in the head and neck region, reports have been made of its occurrence in the intra-oral region [9], nasal bone [10], buccal mucosa [11], tongue [12], parotid gland [3] and jawbones [6]. The maxilla and mandible are somewhat equally affected with five of the previously reported cases affecting the maxilla and the others the mandible. The maxilla was affected in our patient.

Most cases of haemangiopericytoma of the jawbones manifest in the fourth and fifth decades of life. Cases occurring well above 60 years, as in our patient, are however not uncommon as they represent 20% of the previously reported cases. Although the cause of the disease is unknown, an association with trauma has been suggested [14]. The precipitating cause is obscure in our patient, as she did not report any antecedent trauma.

Clinically, haemangiopericytoma presents most often as a slow growing, painless mass and thus medical attention are not usually sought until they grow into large sizes or cause considerable discomfort, usually from pressure on other structures. Our patient had lived with her tumour for at least two years after the recurrence. She sought help mainly because of difficulty she had with nasal breathing and the offensive intra-oral purulent discharge. Pain may be associated with this tumour, especially when it occurs in the bone [15].

The prognosis of haemangiopericytoma is unpredictable, and its benign or malignant nature is not easily discernible histologically, especially in first-time lesion [10] and standard histomorphologic features may be inadequate to predict clinical outcome. This emphasizes the need for long-term follow-up. Its malignant potential is

usually presaged by local recurrence, which occurs in 50% of all lesions subjected to surgical resection. Recurrence, which is known to occur even decades after resection of the initial lesion, is associated with poor prognosis as they may metastasize later. In our patient, we were not conversant with the histological nature of the initial lesion but the fact that it recurred within 5 years of initial resection raised the spectre of malignancy when she reported in our clinic.

In general, all osseous haemangiopericytoma are regarded as malignant *ab initio* although the metastatic potential may vary from one case to the other. Bone haemangiopericytomas are known to metastasize to the lungs, other bones, liver, and local lymph nodes [15]. Metastases were not found in our patient although we did not have the privilege of examining her in the latter phase of her disease and there was no post-mortem examination. Haemangiopericytomas occurring in the abdomen retroperitoneum and the meninges lesions have been reported as being more malignant [16] and are associated with higher local recurrence rates [17] while tumours occurring in the extremities are associated with a longer recurrence-free survival.

Definitive treatment of haemangiopericytoma is primarily surgical. However, surgery is potentially fraught with the danger of severe intractable haemorrhage because of the vascular nature. This was our experience when we carried out the incisional biopsy on our patient. Appropriate planning, wide surgical resection, and long-term postoperative follow-up is associated with better prognosis and has been documented to result in long-term remission or cure [18,19]. Preoperative embolization and afferent vessel ligation may facilitate the surgical resection by reducing the likelihood of potential bleeding [2,6]. It is advisable to identify and evaluate the resected margins of the lesion pathologically.

Radiotherapy after surgical resection also aids remission for incompletely resected masses and it is valuable for inoperable masses. Adjuvant therapies should be considered and evaluated for tumours at sites associated with higher local recurrence rates. In view of the high rate of local recurrence, some workers suggest that the operative sites of all bone lesions should be irradiated [6] while others recommend radiation therapy for positive surgical margins [12]. Chemotherapy has no known role in the management of haemangiopericytoma [6]. However, there is a report on effective control of metastatic disease in a child using combination therapy of actinomycin-D, cyclophosphamide, vincristine and methotrexate[16] and palliative use of Adriamycin- based chemotherapy [12].

The prognosis of haemangiopericytoma remains poor, however, extended survival can be achieved in patients treated with curative intent. Prediction of patient outcome is difficult based on the current knowledge of the biologic behaviour of the disease and histological parameters as local and distant recurrences have been documented after a prolonged disease free interval. Long-term follow up is highly desirable. Potential prognostic factors include tumour grade, size, location, histologic type, resected margins and previous treatment with tumour/histological grade and resected margins being the most important.

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