Craniofacial fibrosarcoma: a case report

JR Moshy[1], HA Mwakyoma[2] and EN Simon[1]

Abstract

Fibrosarcoma (FS) is a malignant proliferation of fibroblasts. Malignancies of fibroblasts are decidedly rare in the oral and oropharyngeal region, but FS is, nevertheless, the commonest mesenchymal cancer of the region, representing more than half of all sarcomas .This article reports a case of a seventy three-years old lady that reported to our clinic with FS affecting the maxilla. The objective of our study was to discuss the biological behavior of this tumour and treatment modalities. Clinical evaluation revealed a huge tumour involving the whole left maxilla. Radiographically the tumour involved both maxillary sinuses and the nose. Histologically, the Haematoxylin and Eosin (H&E) stained tissue sections revealed a poorly differentiated FS from the soft tissue of the maxilla. Surgery, which would entail a wide resection of bones of the region including the floor of the orbit and nasal bones, was thought to be incompatible with life and instead the patient was referred for palliative radiotherapy.

The biological behavior of this tumour (locally destructive, late metastasis) enables our patient to contain that huge and extensive lesion to such an extreme age. As FS may metastasize to the lungs, liver and bones, follow-up of the patient with chest x-rays or CT-Scan would have been very important following surgery. The prognostic factors for FS depend on clinical stage, histological grade of malignancy, tumour location, local recurrences and incomplete resection if surgery contemplated. Taking into account that our case was of a poorly differentiated grade, coupled with the fact that the tumour was located at inaccessible site, factors that pointed to a poor prognosis. These factors affected the survival of the patient with FS.

Key words: Fibrosarcoma, Craniofacial, Biological behaviour, Management.

Introduction

Fibrosarcoma (FS) is a malignant proliferation of fibroblasts. Malignancies of fibroblasts are decidedly rare in the oral and oropharyngeal region, but FS is, nevertheless, the commonest mesenchymal cancer of theregion, representing more than half of all sarcomas. The clinical presentation of FS is varied. The affected age is usually between 30-50 years but there is a wide range and often patients are less than 20 years of age.[2, 10] It can affect both hard and soft tissues.[1] When occurring in bone, the lesion may theoretically arise from theperiosteum, endosteum, or periodontal ligaments and accounts for less than 5% of bone sarcomas.[1] The occurrence of FS in the bone has been reported in association with several conditions namely Paget's disease, fibrous dysplasia and as a post-radiotherapy complication. Fibrosarcoma of bone is a distinctive lesion and should be distinguished carefully from periosteal and soft tissue FS because of differences in prognosis and treatment.[1, 2, 3, 4] About 23% of head and neck sarcomas occur within the oral cavity [2]. Radiotherapy to a local site is known to increase the risk of FS development but there are no other known etiologic factors. On the perioral skin, occasional cases develop at the site of thermal damage or of a pre-existing scar. In some infantile cases, specific translocation (p13; q25) has been described.[5] Craniofacial FS can be classified according to the tissue of origin (soft tissue FS, periosteal FS and intraosseous FS) and histological appearance (well differentiated –low grade, moderate differentiated and poorly differentiated-high grade). The intraosseous FS affects particularly the long bones and its occurrence in the cranium is about 15%, with the mandible being the most common site. The periosteal FS is more prevalent in old age groups and adults. The soft tissue FS is said to have a better prognosis than the introsseous type.[4] Fibrosarcoma is an infiltrative neoplasm that is more of a locally destructive problem than a metastatic problem.[6] This tumour metastasizes through the blood stream rather than lymphatics, thereby producing more wide spread foci of secondary tumour growth [7]. The overall 5-year survival rate ranges between 30 and 50%. Generally, patents with soft tissue FS fare better than patients with primary FS of bone. Also those with well-differentiated FS have a better prognosis than do those with poorly differentiated FS.[6] The treatment of choice is surgical excision with a wide margin. The type of surgical procedure depends mainly on histological grade, local conditions, and tumor location. Well differentiated FS should be treated by wide local excision. Poorly differentiated should be treated by radical surgery including removal of potential invaded muscle and bone. With high probability of metastases after surgical treatment adjuvant modalities should be considered for high grade tumours.[15] We present a case of fibrosarcoma involving the maxilla. The biological behavior and management of this tumour is discussed.

Case report

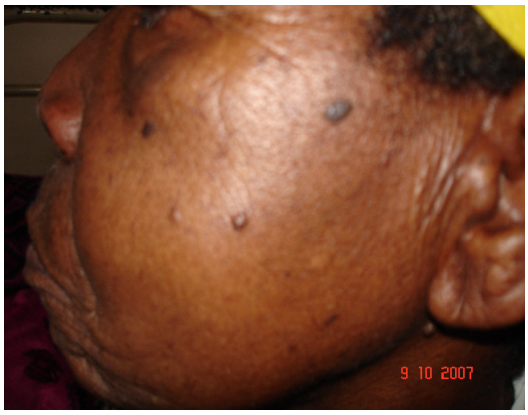
A seventy-three years old lady reported to our clinic with a swelling of the left maxillary region of five months duration. Extra-orally clinical examination revealed a huge tumour involving the whole left maxilla. The overlying skin was normal in colour and texture. Intraorally the tumor involved almost the whole of hard and soft palate (Fig. A&B**).** Radiographically the tumour was found to involve both maxillary sinuses and the nose. The floors of both orbits were not involved. At the onset the tumour was not painful but later it occasionally caused some pain. As the swelling was rapidly increasing in size the pain also simultaneously increased. Except for the pain, the swelling clinically and radiographically resembled any of the fibro-osseous lesions. Histologically, the Haematoxylin and Eosin (H&E) tissue section was lined by a non-keratinized squamous epithelium with partial ulceration in part. Beneath this there was a tumour which was composed of nodular as well as diffuse growth pattern. The cells displayed intertwining fascicles or bundles producing a vague "heringborne" pattern in focal areas. The lesion was typically cellular with minimal amount of mature collagen. The cells were large, pleomorphic with hyperchromatic nuclei and scanty cytoplasm. There were abundant bizarre mitotic figures observed frequently in the tumour. Many blood vessels and haemorrhage were also seen. The histomorphological features were consistent with those of poorly differentiated fibrosarcoma from the soft tissue of

Correspondence to: J moshi,,P. O. Box 650014 Muhimbili University of Health and Allied Sicnes

[1]Department of oral surgery and oral Pathology School of Dentistry, [2]Department of Histopathology/ Morbid Anatomy, MUHAS

left maxilla. (Fig. C, D, E) Surgery, which would entail a wide resection of bones of the region including the orbit and nasal bones, was thought to be incompatible with life and therefore the patient was referred for palliative radiotherapy. She was given a dose of 30 Gray in total for a period of one week. The pain subsided and the tumour shrunk. During the one month follow- up the patient was doing well. However she developed moderate mucositis during the same period of follow-up.

5
A



B

Fig. A. Showing tumour involving Lt upper Fig. B. A firm mass in the left cheeck. The alveolar bone, maxilla & palate swelling was palpable both extraorally and intraorally.

C

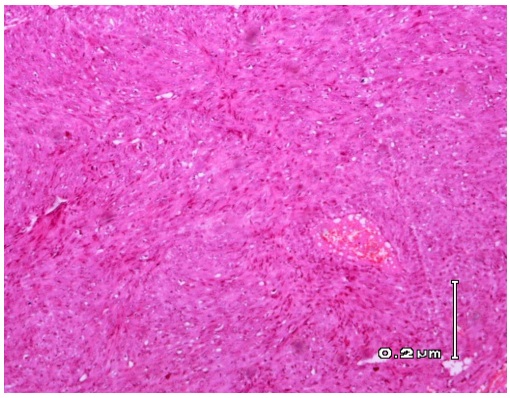
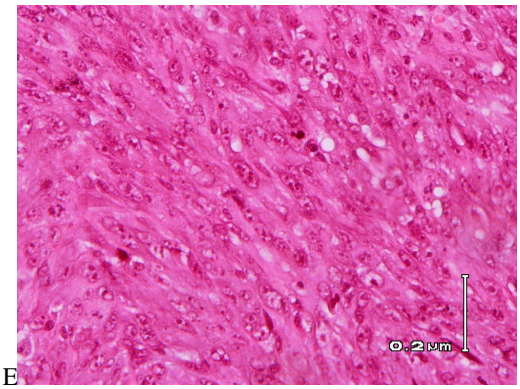
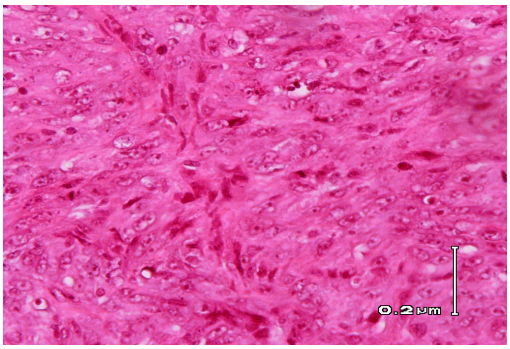


Figure C. Fibrosarcoma showing ill-defined arrangement of the fibroblasts in distinct intersecting fascicles (X 10) 6`

D



E



Figures D&C:Fibrosarcoma, the spindle cells show considerable nuclear variation (D) with numerous mitotic figures (E){ X 40}

Discussion

The clinical presentation of FS is varied. The affected age is usually between 30-50 years but there is a wide range and often patients are less than 20 years of age.[2, 10] Our case was aged 73 years, the age which is different from the peak age reported in the literature. It has been reported in the literature that FS is an infiltrative neoplasm that is more of a locally destructive problem than a metastatic problem.[4, 6, 7] It is seldom metastasizes except late in its clinical course. The biological behavior of this tumor (locally destructive problem, late metastasis) might have enabled our patient to contain that huge and extensive lesion to such an extreme age.

Intraorally, FS may arise in any location but it commonly involves the periosteum of the maxilla or mandible.[10] However, FS may arise in a scar tissue sometimes many years after the scar developed as a result of cellular instability from chronic inflammation. Fibroblasts are capable of a wide variety of phenotypic modulations in response to different injuries.[3, 4] Fibroblasts are very adaptive and may alter their phenotype in response to environmental changes. Such modulations are reflected in architectural, ultrastructural and immunoreactivity. The immunophenotypic variations usually reflect morphological functional states and also lead to diagnosis confusion. Larson et al. reported that periosteal FS is more prevalent in old age groups and adults. Our case was aged 73 years and the lesion was located in the maxilla. It follows therefore that; the FS in present case might have arisen from the periosteum of the

maxilla and extended locally to involve the maxillary bone and other contiguous midfacial bones. On the other hand, the occurrence of FS in the bone denotes that the lesion may theoretically arise from periosteum, endosteum or periodontal ligaments.(6)

FS clinically presents as an innocuous, lobulated, sessile, painless and non-hemorrhagic mass of normal coloration. FS may present as a rapidly enlarging hemorrhagic mass similar in clinical appearance to an ulcerated pyogenic granuloma, peripheral giant cell granuloma or peripheral ossifying fibroma. Even lesions that do not demonstrate surface ulceration or rapid growth may show destruction of underlying muscle and bone. Intraosseous FS may also present as a swelling associated with pain.(1,3) Our patient presented with pain associated with the tumour. Usually primary intraosseous FS is asymptomatic but in about 30% of cases it may present with some symptoms.(3, 9) The association of pain in the current case adds a possibility that this case might have aroused from the endosteum and that it is possible that it was a primary intraosseous fibrosarcoma.

FS is a lesion with varied microscopic appearance. The low grade (well differentiated) variant is usually somewhat circumscribed and composed of mature spindle cells that differentiate from benign fibrous hyperplasia. The presence of focal area of anaplasia and increased mitotic activity in such cases indicates aggressive behavior. Cells display intertwining fascicles or bundles producing a" herringbone"parttern in focal areas. The lesion is typically cellular but moderate amounts of immature collagen may be produced sometimes with areas of hyalinization. Cells and nuclei are not pleomorphic but have scattered normal mitotic figures. The less well differentiated (moderately differentiated) has minimal collagen production and marked cellularity. Cells are larger and more hyprechromatic. There is more pleomorphic and more round nuclei. Multinucleated giant cells are rarely seen. The lesion is mostly observed in young patients (2). Poorly differentiated lesions have focal areas of tumor necrosis and myxoid areas. However, there is no stromal hemorrhage. In adult FS is extremely rare in the mouth and that the diagnosis is many times of exclusion in a lesion which is negative to appropriate immunohistochemical markers. FS contains cells which react positive for vimentin only.(3, 8, 11, 12, 13) Our present case was diagnosed as high grade (poorly differentiated) FS.

The management of craniofacial FS involves detailed clinical, radiological and histopathologic evaluation. The combination of these features would lead to a proper diagnosis. Depending on the location in the craniofacial region, two views of plane x-rays should be taken (perpendicular to each other) to confirm presence of the tumour followed by superior radiological modalities to determine the extent of the lesion and for proper planning of the surgical approach. Such radiological evaluation should include orhopantomography (OPG), poster anterior view of the skull (PA-skull), chest X-ray (to rule out distant metastasis) and where possible CT/ MRI (For extension & planning of surgery). FS radiographically appears as lytic lesion with geographic moth eaten or permeative bone destruction. The cortex is thinned or destroyed and soft tissue extension occurred in more than 80% of cases.

The treatment of choice is surgical excision with a wide margin. The type of surgical procedure depends mainly on histological grade, local conditions, and tumor location. Well differentiated FS should be treated by wide local excision. Poorly differentiated should be treated by radical surgery including removal of potential invaded muscle and bone. With high probability of metastases after surgical treatment adjuvant modalities should be considered for high grade tumours. However, the need for adjuvant radiotherapy or chemotherapy is still controversial.(3) Radiotherapy may be used as salvage for recurrences and in cases with high grade of malignancy.(15) With FS that occurs in the facial region treatment becomes quite challenging because of the anatomical limitations as it was the case in our patient. Reconstruction in the form of flap and reconstruction plate should be considered to close the defect left behind following surgery. Prophylactic neck dissection is still controversial.(5) but may be considered in high grade tumours because these tumours may present subclinical or microscopic metastases at the time of diagnosis. Following treatment, the five year survival is poor ranging from 20-35% (2). As FS may metastasize to the lungs, liver and bones follow up of the patient with plain chest x-ray or CT-scan is very important after surgery. These patients need to be followed up for long period.The prognostic factors for FS depend on clinical stage, histological grade of malignancy, local recurrences and tumor location.(5) These factors affect the survival of the patient with FS. The higher the grade or stage the poor the prognosis. Also incomplete resection of the tumor is associated with high recurrences and hence poor prognosis. However, the soft tissue FS has a good prognosis compared to intraosseous tumors.

Conclusion and Recommendations

Our patient could not be treated by surgery because of late reporting when the tumour had spread to the extent that surgery was thought to incompatible with life. In Tanzania patient start to present their problems at dispensary level where they are referred to a district level and later to regional hospitals. It is from here that they can be referred to any of the four referral centers in the country. In the case of maxillofacial pathologies, the only center the patients can get adequate management is the Muhimbili National Hospital. It is not easy for the majority of patients who belong to low socio-economic group to follow this long process, hence the delay. This emphasizes the need for raising the awareness of the general public and health personnel at primary centers to make accurate, provisional diagnosis of any maxillofacial tumour and make early referrals to appropriate centers for accurate diagnosis and timely management.

References

1. **Papaglopoulos P.J , Galanis E.C , Trantafyllidis P , Bscainois P.J , Sim F.H , Unni K.K. Clinicopathologic features, diagnosis, and treatment of fibrosarcoma of bone. Ann. J. Orthop. 2002; 31: 253-257.**
2. **http// www. Maxillofacial centre. com./ Bond Book/ Soft tissue / fibrosarcoma.**
3. **C.M. Pereira, J.Jorge Jr, O.Di Hipolito Jr , L.P. Kowalski, M.A. Lopes . Primary intraosseous fibrosarcoma of jaw. Int. J. Oral Maxifac. Surg. 2005; 34: 579-581.**

4. **Larsson S.E., Lorentzon R., Boquist L. Fibrosarcoma of bone. A demographic, clinical, histopathological study of all cases recorded in the Swedish cancer registry from 1958- 1968. J. Joint Surg. Br. 1976; 58: 412-417.**
5. **Knezevich SR, Mcfadden DE, Tao W, Lim JF, Sorensen PH. A novel ETV6-NTRK3 gene fusion in congenital fibrosarcoma. Nat Genet 1998;18:184-187.**
6. **Regezi JA, Scuibba JJ. Fibrosarcoma. In Oral Pathology; clinical pathologic correlations, 3rd ed. W.B. Saunders Company, Philadelphia 1989, 191-192.**
7. **Shafer GW, Hine MK, Levy BM. A text book of Oral Pathology, 4th ed. W.B. Saunders, Philadelphia 1983, 169-171.**
8. **S. Wada, L. Yue, I. Furuta and T. Takazakura. Leiyomyosarcoma in the maxilla. A case report. Int. J. Maxillofac. Surg. 2002; 31: 219-221.**
9. **Dry S.M., Jorgensen J. L., Fletcher C. D. M. Leiomyosarcoma of the oral cavity. An Unusual Topographic Subset easily mistaken for non mesenchymal tumors. Histopathology 2000; 36: 210-220.**
10. **Stanley Kerpel, Paul Freedman, Stephen Troyer. Expansile mass of the body of the mandible. J. Oral Maxillofac. Surg. 1992; 50: 627-632.**
11. **Greager J. A., Reichard K., Campara J. P. Fibrosarcoma of the head and neck. Am. J. Surg. 1994; 167: 437-439.**
12. **Antonascu C. R., Erlandson R. A. Fibrosarcoma mimicking plasmacytoma or carcinoma. an ultrastructural study of 4 cases. Ultrastruct. Pathol. 2001; 25: 31-37.**
13. **Slootweg P. J., Roholl P. J., Muller H. et tal. Spindle- cell carcinoma of the larynx. Immunohistochemical aspects. J. Cranio-maxillofac. Surg. 1989; 17: 234-236.**
14. **Berton F., Capanna R., Clderoni P. Primary central (medullary) fibrosarcoma of the bone. Semin Diagn Pathol 1984; 1: 185-198.**
15. **ndberg RD, Martin RG, Romsdahl MM. Surgery and postoperative radiotherapy in the treatment of soft tissue sarcomas in adults. AJR 1975; 123:123-129.**