FIBROSARCOMA OF MAXILLA - A RARE PATHOLOGIC ENTITY

Prabu Vijay*, Kannan Suresh, John Reena*****

Fibrosarcomas of the oral and maxillofacial region is a rare entity with a poor prognosis. Only 10% of fibrosarcomas occur in head and neck region. The incidence of fibrosarcoma is less than other malignant mesenchymal lesions such as liposarcoma, rabdomyosarcoma, and malignant fibrous histiocytoma. Fibrosarcoma arising in sino-nasal cavities are rare and not diagnosed early till they reach a size of enormous proportion. The reason being the anatomy of maxilla which allows the growth to silently occupy the air sinuses such as maxillary sinus and ethmoidal sinuses without any symptoms. Hence the inability to provide surgical margins free of disease results in reccurence.We report a case of fibrosarcoma maxilla involving the maxillary sinus in a 33years old female patient.

Key words: Fibrosarcoma, maxilla tumors, maxillary sinus, sino-nasal tumors.

Fibrosarcomas are defined as malignant tumors of fibroblast that show no other evidence of cellular differentiation and are capable of recurrence or metastasis. Based on the histology they are often graded with a grading scale of 1 to 3 [13].

Fibrosarcomas in the maxillofacial area arise from bone, periosteum, or muscle fascia.Intraoral fibrosarcomas may arise commonly in the cheek, maxillary sinus, pharynx, palate, lip and periosteum of maxilla and mandible [13].

Fibrosarcoma show a slight female predilection and occur predominantly among young adults. They show a greater tendency to metastasize through blood stream thereby producing more widespread foci of secondary tumor [12].

Oral fibrosarcomas are locally infiltrative and destructive. Metastasis by the lymphogenous route to the local lymph nodes or by haematogenous route to the lungs, brain or bone is also reported [12]. Surgical management of fibrosarcoma in maxillofacial region is far from satisfactory, because of lack of inadequate clearance. Like any soft tissue sarcoma, surgery should be followed by adjuvant radiotherapy [12, 8].

* *Post graduate student*
** *Reader*
*** *Professor and Head of the Department, Dept of Oral surgery, Vinayaka Missions Sankarachariyar Dental College, Ariyanoor, Salem, Tamil Nadu.*

CASE REPORT

A 33 years old female reported to the Dept of oral and maxillofacial surgery with complaint of swelling in the left side of the face and a growth inside the mouth of six months duration. Her past dental history is relevant for having undergone extraction of a mobile upper left tooth following which the swelling increased in size.

Extra oral inspection revealed a diffuse swelling in the left side cheek measuring approximately 2.5 x 2 cm, extending from the infraorbital rim to angle of the mouth (superoinferiorly) and from the nasolabialfold to the malar region (anteroposteriorly). On palpation swelling was firm and nontender.

Intraoral examination revealed a pinkish-red growth extending from left canine to second molar region (anteroposteriorly) obliterating the buccal sulcus (superiorly)and medially up to 1cm away from the midline of palate. The surface of the swelling was ulcerated which could be due to trauma from opposing tooth. On palpation the swelling was nontender, soft to firm in consistency.

Orthopantamogram and CT scan revealed an ill-defined osteolytic lesion from canine to second molar region and extending into the palate. Reports of routine blood, urine and chest X-ray were within normal limits. Histopathological examination of incisional biopsy revealed numerous spindle shaped fibroblast

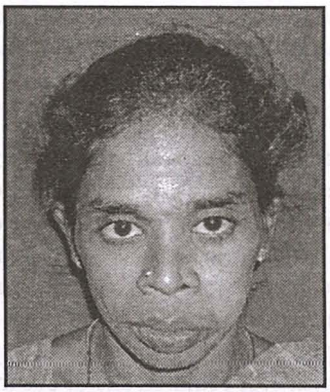


Fig. 1: Pre-op Front view

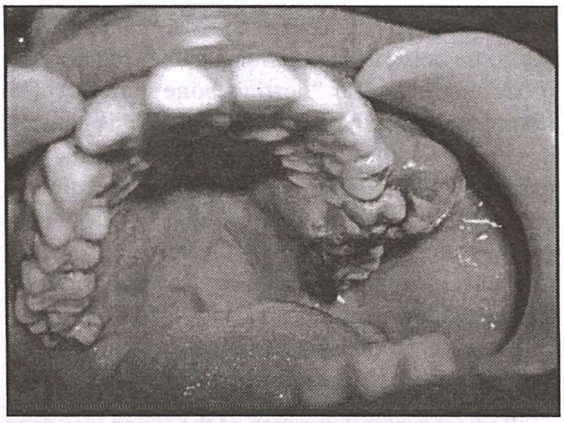


Fig. 2: Pre-op Intra oral lesion

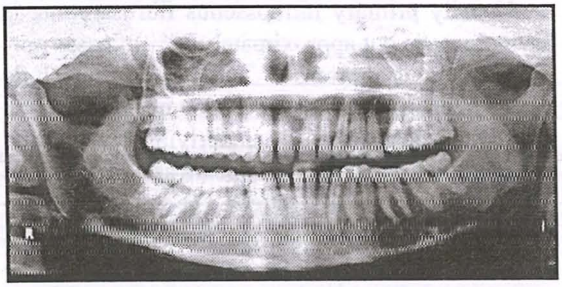


Fig. 3: OPG showing radiolucent lesion in 24, 25, 26 region

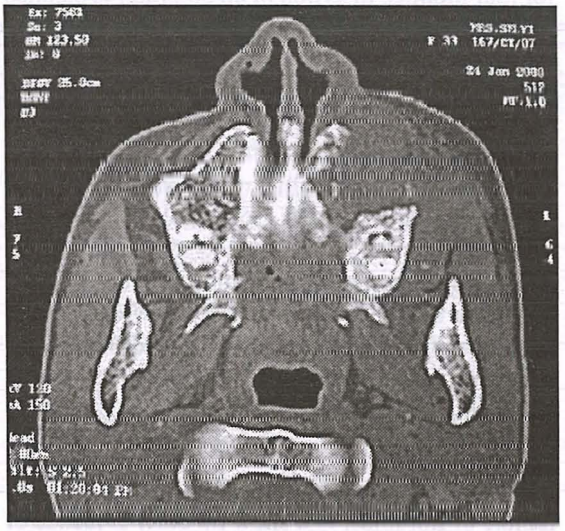


Fig. 4: Axial CT showing erosion of maxillary alveolus and palate



Fig. 5: Weber-Fergusson incision

cells within the connective tissue stroma proliferating in streaming fashion forming interlacing fascicles producing classical "herring bone pattern". The fibroblast cells exhibited increased mitotic activity, hypercellularity, cellular pleomorphism and hyperchromatism. In several cells the chromatin material was irregularly dispersed throughout the nucleus, while in others it was condensed and stained intensely. Immunohistochemistry showed positive for staining with vimentin and negative for desmin, actin, epithelial membrane antigen. The specimen was reported as aggressive fibrosarcoma.

Radical surgical excision of the lesion was done (total maxillectomy including the orbital floor). The margins of the specimen were positive for the disease. Hence the patient was subjected to postoperative radiotherapy for a period of six weeks. A total of 6000cGy of radiation was given to patient in divided doses. Her wound healing and response to radiotherapy was satisfactory. She has been followed up for four months. Insertion of an obturator is planned to aid in feeding and deglutition.

DISCUSSION

Fibrosarcoma of the head and neck region is rare and the incidence is 0.4% to 1% of all soft tissue sarcomas [1, 3, 9]. This entity was first described by Christoperson [13].Between 1950 and 1975; they were believed to be the most common soft tissue malignancy. However, the identification and separation of malignant fibrous histiocytoma, fibromatoses, nodular fascitis, fibrous osteosarcoma and undifferentiated epithelial malignancies, all of which were previously regarded as fibrosarcomas has unmasked the rarity of true fibrosarcoma [13].

Today fibrosarcoma are defined as malignant tumors of fibroblast that show no other evidence of cellular differentiation and are capable of recurrence and metastasis. They are often graded with grading scale of 1 to 3. Grade 1 and 2 fibrosarcoma tend to be more differentiated and therefore less clinically aggressive. Grade 3 fibrosarcomas are usually faster growing and very aggressive; they also have a greater tendency to metastasize [13].

Fibrosarcomas are predominantly found in adolescent and young adults with higher incidence in young females. The common site of involvement are thigh (68%), upper limb (26%), abdominal wall (4%), and rarely in head and neck area (<1%) [8, 10].

Fibrosarcoma of the head and neck region is well documented without site predliction.23% of head and neck fibrosarcomas occur within the oral cavity. On the perioral skin, occasional cases develop at the site of thermal damage or of a pre-exisisting scar. Spector reviewed all head and neck tumors in his series and found only 12 cases of head and neck fibrosarcoma.Most common site of occurrence was reported as base of the tongue [14].Chatterji reported one case of fibrosarcoma of Nasal cavity and Para nasal sinuses [3].

Usually primary intraosseous fibrosarcoma is asymptomatic but approximately 30% of cases may present some symptoms [2, 4]. Even lesions which do not demonstrate surface ulceration or rapid growth may show destruction of underlying muscle and bone. Radio graphically it appears as a purely lytic lesion with a destructive pattern. The cortex is thinned or disrupted and soft tissue invasion is detected in up to 86% of cases [4, 16]. In our case, the patient reported with a fairly big lesion and large area of bone destruction was also evident radiographically. The lesion had rapid growth rate, which was seen between the period of incision biopsy and surgery, which indicates the aggressiveness of the same.

The treatment of fibrosarcoma is radical surgical removal with 3 to 5cm clearance margins. This is not possible in the oral cavity and sino-nasal region because of anatomy and mutilation effect. This restriction leads to failure of local control which results in recurrence [6, 15].

In a series of head and neck fibrosarcomas studied by Liberman over 63years it was concluded that it is difficult to gain meaningful therapeutic statistics with such a rare tumor [8].The overall survival rate at 10 years may vary from 21.8% to 83% and clinical stage, histological grade of malignancy and local recurrences are the most important prognostic factor [2,



Fig. 6: Total maxillectomy done

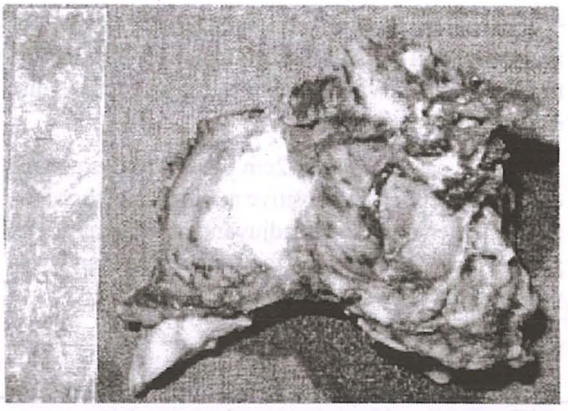


Fig. 7: Excised lesion

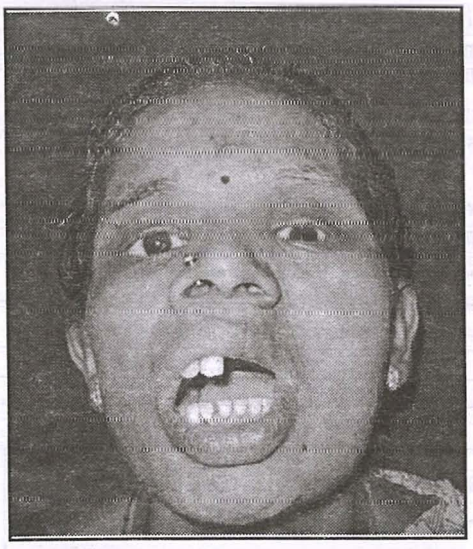


Fig. 8: 4 months Post-op Front view

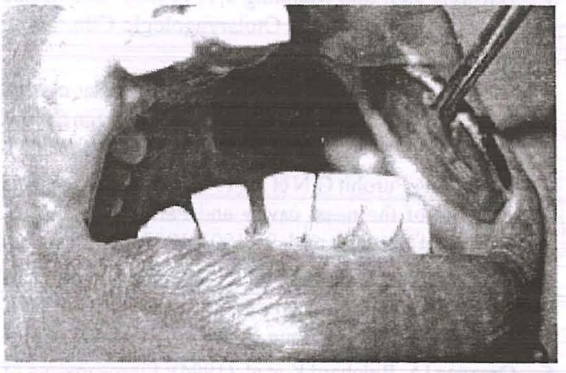


Fig. 9: 4 months post-op Intraoral view

[5]. The role of adjuvant radiotherapy and or chemotherapy is still unclear but is indicated in high grade tumors, lesions with close or positive margins and size larger than 5cm [5,15].

In our case inspite of 2cm clearance margin the excised specimen had positive margins in all borders. So the patient underwent adjuvant radiotherapy. The patient was followed up for four months and was without any clinical evidence of recurrence.

SUMMARY

Fibrosarcoma arising from the oro-naso-sinus region are extremely uncommon tumors that are usually in the advanced stage when diagnosed. Its location preludes wide surgical margins, which generally results in recurrence and poor prognosis. Even with adjuvant radiotherapy, the recurrence rate is high. Inclusion of neo-adjuvant chemotherapy in modern regimes may facilitate resection of unresectable tumors and improve the prognosis for these patients.

BIBLIOGRAPHY

1. Batsakis J.G. and Manning J.T. (1986): Soft tissue tumors: unusual forms, Otolaryngologic Clin. North Am, 19(4). 659-683.
2. Bertoni F, Capnna R et al. (1984): Primary central (medullary) fibrosarcoma of bone, Semin.Diagn pathology, 1:185-198.
3. Chaterji.P, Purohit G.N et al.(1977): Alveolar soft part sarcoma of the nasal cavity and Para nasal sinuses, Journal of Laryngo otol, 91;1003-1008.
4. Eyre-Brook AL, Price CHG. (1969): Fibrosarcoma of bone. Review of fifty consecutive cases from the Bristol tumor registry, J Bone Joint Surg, 51B:20-37.
5. Greager JA, Reichard K et al. (1994): Fibrosarcoma of head and neck, Am J Surg, 167:437-439.
6. Hamid Mansouri et al.(2006): Fibrosarcoma of the maxillary sinus, Indian journal of otolaryngology and Head and Neck surgery, 58(1):104-105.
7. Huvos AG, Higinbotham NL.(1975): Primary fibrosarcoma of bone. A clinicopathologic study of 130 patients, Cancer ,35:837-847.
8. Liberman P.H, Brennan.M.F et al. (1989): Soft part sarcoma: A clinicopathologic study of half a century, Cancer, 63:1-13.
9. Mark R.J, Sercaz J.A et al. (1991): Fibrosarcoma of the head and neck: The UCLA experience, Arch otolaryngol Head and Neck surg, 117:396-401.
10. Meis Kindbolm J.M, Kindbolm L.G et al. (1995): Sclerosing epitheloid fibrosarcoma: a rare variant of fibrosarcoma simulating carcinoma, Am. J. Surg. Pathology, 19: 979-993.
11. Neville BW, Damm DD et al.(1995): Oral and maxillofacial pathology.(1st edn.), W.B.Saunders company ,Ch 4: 469
12. Rajasekhar Halli, Manjula Hebbale. (2007): Fibrosarcoma- A rarity in Maxillofacial region, Jr of Maxillofacial and oral surgery, 6(1): Pg 34-36.
13. Robert E. Marx, Diane Stern. (2003): Oral and Maxillofacial pathology: A rationale for diagnosis and treatment, Quintassence publishing Co Inc Illinois, Pg: 466-468.
14. Spector R A, Travis L.W et al. (1979): J Alveolar soft part sarcoma of head and neck, The Laryngoscope, 89: 1301-1306.
15. Rockley TJ, Liu KC. (1986): Fibrosarcoma of the nose and paranasal sinuses, Laryng Otol, 100:1417-1420.
16. Taconis WK et al. (1985): Fibrosarcoma of long bones: A study of the significance of areas of malignant fibrous histiocytoma, Br Jr of Bone joint surg, 65:111-116.

Our mission as Fellows in the Pierre Fauchard Academy is to consistently focus on professionalism, integrity and ethics worldwide, by our own conduct as worthy role models, by the advancement of dentistry to the highest level, by supporting and honoring colleagues for their distinguished work, research, contributions, and public service, and by providing excellence in programs, education and example.