Malignant Fibrous Histiocytoma of Maxilla – A Report of Five Cases and Review of Literature

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Malignant fibrous histiocytoma of the maxilla is a rare malignant tumor of the bone. Affection of maxilla does not occur very commonly. In this report five cases of fibrous histiocytoma have been presented along with a review of literature.

INTRODUCTION

Malignant fibrous histiocytoma (MFH) is a pleomorphic sarcoma composed of both fibroblasts and histiocytes like cells arranged in a storiform pattern. The tumor was first described by O'Brien and Stout in 1964. The histogenesis of this tumor remains controversial. Earlier electron microscopic studies suggested that the precursor cells, which by means of bimodal differentiation gave rise to histiocytes and fibroblasts (Fu 1975). More recent studies however suggest that myofibroblasts may be the cell of origin (Churg 1977, Hashimoto 1982). A review of English literature revealed only 20 cases of histologically diagnosed malignant fibrous histiocytoma of maxilla. In this report we discuss the clinical presentation, treatment policy and follow up results of 25 cases of MFH (including 5 cases of present series).

MATERIAL AND METHODS

The clinical records of five new cases of malignant fibrous histiocytoma of maxilla were reviewed. The patients were treated at Nehru Hospital, Post Graduate Institute of Medical Education and Research, Chandigarh, from 1985 to 1995. The clinical chart of each patient was reviewed for the following data. Age at diagnosis, sex, initial symptoms, duration of symptoms, site of origin, radiological findings, histological type, evidence of metastasis either at diagnosis or

during course of treatment, initial treatment and status of disease at last follow up. Follow up was obtained through clinical records and direct correspondence with the patient or a family member.

OBSERVATIONS

Age and Sex : The majority of cases of maxillary MFH occur in 5th to 7th decade of life (age range 10-79 years). There is male preponderance in the ratio of 2.4:1.

Symptoms : The commonest symptoms noted were cheek and palatal swelling, loose teeth and dental pain, facial pain, infraorbital nerve anaesthesia, purulent discharge from nose and epistaxis. Table I shows the frequency of occurrence of these symptoms. The duration of initial symptoms in these patients varied from three months to a year.

Table I
Showing the frequency of Symptoms in cases of MFH

S. No.	Symptoms	No. of Cases	% of Total
1.	Facial swelling	16	66.6
2.	Facial pain	12	50
3.	Dental problems	07	29.1
4.	Palatal swelling	06	25
5.	Infraorbital anesth	06	25
6.	Purulent nasal dis.	05	21.1
7.	Epistaxis	02	8.3
8.	Trismus	01	4.1
9.	Proptosis	01	4.1

*Duration of symptoms 3 months to 12 months

Radiography

The commonest radiographic finding seen on plain x-ray was a bony erosion. The other radiological features seen were opacity of maxillary sinus, periapical radiolucency and lytic area in the maxilla with a soft tissue mass. In 4 of our 5 cases CT scan was done. This investigation helped us to know the exact extent of tumor preoperatively.

Histopathology

23 out of 25 cases of MFH of maxilla showed storiform pattern on histopathology. One patient had myxoid and one pleomorphic variety. Our five cases had storiform variety.

Treatment and Follow up

Treatment included surgery only, surgery combined with radiotherapy either preoperative or postoperative, surgery combined with both radiotherapy and chemotherapy, radiotherapy alone and radiotherapy in combination with chemotherapy. The preferred mode by majority of authors is surgery in combination with radiotherapy (14/25) or surgery alone (8/25) (Fig.1).

Fig.1

Showing the treatment modality in MFH cases

Recurrence rate at the local site was 32% (8/25) within a period of 1 year from diagnosis. The common sites of local recurrence noted were skull base, orbit and maxilla. Treatment response of the recurrence was poor in the skull base

region (2 cases). Both of them died within 1 year from the initial diagnosis.

Distant metastasis was reported in 8 cases (33.3%). The common sites were lungs, mandible, scalp, brain, etc. Mortality rate as available from 1 year follow up data of each case is 25% (6/25). Of these 6 cases who died, 1 case had metastasis in the lungs and lymph node, 1 case in the lungs and skull base, 1 to the liver, lumbar spine and calvarium, 1 to the lymph nodes only and 1 at skull base.

CASE REPORT

Case 1, CS, a 31 years male presented with swelling right cheek of 12 months duration and another swelling at right upper alveolus following tooth extraction of 3 weeks duration. Examination revealed a firm subcutaneous swelling over the cheek between the body of zygoma and the nasooptic and alarfacial grooves. The infraorbital bony margin was intact but the lower conjunctival fornix showed fullness. There was a reddish firm lobulated mass replacing the premolars and molars. X-ray paranasal sinuses showed a diffuse opacity in the maxillary sinus with destruction of anterolateral, medial and inferior walls. Biopsy of lesion showed it to be Malignant fibrous histiocytoma (Fig.2). Total maxillectomy was performed with preservation of eye ball. Histopathology was storiform

Fig.2

H & E x 140 showing MFH with storiform pattern

Malignant fibrous histiocytoma. Postoperative radiotherapy in a dose of 55 Gy was given to the patient. 5 months later an orbital exenteration had to be performed for local recurrence. Patient is free of disease 2 years after treatment.

Case 2

SR, 62 years male was admitted to ENT ward with complaints of epistaxis, mass coming out of the nasal cavity and nasal obstruction of 4 months duration. There was no history of decreased vision, E.N.T examination revealed proptosis of right eye and a fleshy mass filling the right nasal cavity. Posterior rhinoscopy showed the mass occupying the right choana. Visual acuity was 6/12 in the right eye and 6/9 in the left eye. CT scan revealed bony destruction of maxilla and invasion of orbit and infratemporal fossa (Fig.3).

Fig.3

CT Scan showing invasion of orbit and infratemporal fossa

Biopsy of the tumor showed it to be Malignant fibrous histiocytoma. Total maxillectomy was performed sparing the eyeball as patient did not agree for orbital exenteration. Post operative radiotherapy was given to the patient in a dose of 55 Gy. Patient had residual disease on 9 months follow up.

Case 3

S. K., 48 years female came with history of excruciating facial pain and difficult in opening of mouth and swelling left cheek of 3 months

duration. ENT examination revealed trismus and ill defined swelling in the cheek corresponding to the level of 3rd molar of upper jaw. Infraorbital rim was intact. CT scan revealed bony destruction of posterolateral wall of maxilla and a soft tissue mass occupying pterygopalatine and infratemporal fossa. Total maxillectomy along with clearance of tumor from infratemporal fossa was done. She developed local recurrence at skull base and distant metastasis in the lungs 6 months after the diagnosis. She died of the disease 8 months after surgery.

Case 4

RK, 36 years male presented with complaints of left nasal obstruction, purulent nasal discharge and epistaxis of 5 months duration. ENT examination revealed a reddish mass occupying the floor and middle meatus of left nasal cavity. Posterior rhinoscopy was normal. CT scan showed destruction of medial wall of maxillary sinus and soft tissue mass occupying left maxillary antrum and nasal cavity. Total maxillectomy was performed under general anaesthesia. Patient is alive with no evidence of disease after diagnosis.

Case 5

DK, 42 years male was admitted with complaints of swelling right cheek and palate of 4 months duration. Swelling in the cheek was extending from body of zygoma to nasooptic and alar facial groove. The swelling was nontender, smooth and firm in consistency. Infraorbital margin was intact. There was another swelling occupying the premolar and molar region of maxilla. CT scan revealed destruction of anterolateral and medial walls of maxilla. Infratemporal fossa and orbit was free. Total maxillectomy was performed. Patient is alive with no evidence of disease 4 years after diagnosis.

DISCUSSION

Malignant fibrous histiocytoma is now recognized as the most common soft tissue sarcoma in adults (Enzinger 1986, Wiess et. al. 1978). About 10% of Malignant fibrous histiocytoma occur in the head and neck region.

(Weiss et al. 1978, Huvois et al. 1985, Russel et al. 1985, Enjoji et al. 1980) with sinonasal tract as most common location accounting for 30% of all cases (Blitzer 1981). This neoplasm has also been described to occur after irradiation for ameloblastoma (Li 1977). Majority of cases of epidermoid carcinoma of maxilla occur in 3rd to 5th decade of life, while malignant fibrous histiocytoma cases present usually between 5th to 7th decade of life. In the present report 16 cases (64%) were in 5th to 7th decade of life.

The diagnostic feature of MFH on histopathology is presence of storiform fibrous stromal background. Weiss et al. 1978 identified three patterns in MFH; Storiform, Pleomorphic and fascicular. Ogura et. al (1980) divided MFH into four categories. Storiform predominance, myxoid predominance, inflammatory predominance and pleomorphic. This classification has some significance with respect to prognosis. Storiform and myxoid patterns are associated with good prognosis as they metastasize slowly and respond well to surgical therapy. The inflammatory and pleomorphic variants are aggressive and metastasize early and respond less favourably to surgical excision. However there are no reliable histologic criteria for predicting the metastatic or recurrent malignant potential of an individual tumor. Nevertheless Soule and Enrique (1972) contended that an anaplastic appearance of stromal cells in fibrous histiocytoma indicated malignancy.

Treatment usually consists of wide local excision with adjuvant irradiation reserved for patients with close or positive surgical margins and chemotherapy for metastasis. Dahlin (1981)

believes that it should be treated like fibrosarcoma with surgical ablation. It is difficult to come to a consensus for the treatment policy of maxillary Malignant fibrous histiocytoma as results of various modalities of treatment have been difficult to assess due to paucity of data. In this series of 25 cases, 23 patients have undergone surgical ablation without or with combination of radiotherapy or chemotherapy. Few authors (Blitzer 1981, Barnes 1985) have reported that approximately 20-40% patients with Malignant fibrous histiocytoma will experience local recurrences, majority within the first year of treatment. In this series of 25 patients, 9 patients finally succumbed to the disease, 6 of them in the first year of diagnosis. Block et. al. (1986) reviewed 15 cases of Malignant fibrous histiocytoma of maxilla and reported one of their own cases. Metastatic lesions or extensive local recurrences occurred in 9 of the 16 patients after surgery and radiation therapy. Histopathologically 14 lesions were classified as storiform, one as myxoid and one as pleomorphic. Barnes et al. (1988) in a report of 12 cases of Malignant fibrous histiocytoma of head and neck had 4 cases in the sinonasal tract. 3 of their cases had local recurrence with eventual death. One patient was alive with no evidence of disease after 8 months of treatment.

To conclude, malignant fibrous histiocytoma is an aggressive neoplasm which may originate anew or may arise as a complication of irradiation or some other underlying condition. Total surgical margins may bring hope to a patient with this neoplasm as otherwise recurrence are quite common.

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