Malignant Fibrous Histiocytoma of the Maxilla

— Report of A Case —

Yoon Kyeong Oh, M.D.[1] and Hwan Ho Yeo, D.D.S.[2]

Department of Therapeutic Radiology[1], Chosun University Medical College,
Department of Oral & Maxillofacial Surgery[2], Chosun University Dental College, Kwangju, Korea

= Abstract =

Malignant fibrous histiocytoma(MFH) of the maxilla is a rare malignant bone tumor. Seven percents of all MFH occur in the head and neck. Approximately 12% of these tumors occur in the maxilla.

Local recurrence or distant metastasis was reported in 55% of cases of maxillary MFH. The mean survival time of 30 months was reported from a review of 14 MFHs in the maxilla, mandible and oral soft tissues.

MFH of the maxilla is best treated surgically but radical neck dissection does not appear to be indicated unless there is clinical evidence of lymph node metastases. Although the use of radiation therapy for head and neck MFH has not been studied for a series of cases, individual cases of regression or histological change have been reported. Other authors have reported numbers of cases who received radiation therapy without benefit. Response to combination chemotherapy has been reported in 33% of 23 patients with recurrent or metastatic MFH.

We report here a case of MFH occurring in the maxilla with a review of literature about the clinical behavior and treatment of these lesions.

Key Words : Malignant Fibrous Histiocytoma, Radiation Therapy, Maxilla

INTRODUCTION

Malignant fibrous histiocytoma (MFH) is now the most frequently diagnosed soft tissue sarcoma in adults. In a recent Scandinavian histopathology review of high-grade soft tissue sarcomas, 40% of the cases were MFH, followed by synovial sarcoma (15%), leiomyosarcoma (9%), liposarcoma (8%), and malignant schwannoma (6%)[1-2]. Malignant fibrous histiocytoma occurs mainly in the 5-7th decades, with a reported age range of 6-89 years[3].

Although MFH is now recognized as the most common soft tissue sarcoma of late adult life, it rarely occurs in bone and then more usually in the long bones. Presentation affecting the facial bones and skull is extremely rare (up to 17%), but MFH should be considered in the differential diagnosis of bone tumors affecting the maxilla. Skeletal lesions appear more aggressive than those in soft tissues. Early radical excision is the treatment of choice although radiation therapy for inoperable tumors may lead to regression[4].

Pezzi et al. examined the prognostic factors in 227 patients with MFH. Grade and size emerged as significant prognostic indicators[5]. In other study about postoperative radiotherapy for MFH the status of surgical margins may influence the outcome of postoperative radiotherapy for MFH[1].

Also analysis of local recurrence rate by site revealed 3 of 4 (75%) in the head and neck, 3 of 8 (38%) in the trunk, 3 of 8 (38%) in the pelvis, and 4 of 29 (14%) in the extremities[1].

Weiss and Enzinger reported that 7% of all MFH occur in the head and neck. Approximately 12% of these tumors occur in the maxilla[6-7]. Blitzer et al. showed the maxilla was the most common single intrabony site of presentation of this tumor[7]. The first case of MFH affecting the maxilla was reported by Feldman and Norman in 1972[8], and a further 16 cases have been added to the literature[9].

The lesion consists of fibroblasts and histiocyte-type cells, often arranged in a storiform pattern[1, 3]. Histiocytes may also be present as multinucleated giant cells and cells with a foamy cytoplasm. There can be extensive cellular pleomorphism and many tumor giant cells. A wide variation in the number of mitoses has been observed. Based on histological features, MFH has been divided into five histological types. The storiform-pleomorphic variant is the most common and other types include: myxoid, giant cell, inflammatory and angiomatoid. The significance of this classification may be important with respect to prognosis: tumors with angiomatoid and myxoid patterns are often associated with a more favorable prognosis as their metastases occur late and respond well to surgery[6]. The inflammatory and, to a lesser extent, the pleomorphic types are however more aggressive, metastasize early and respond less favorably to surgery alone[9].

We report here a case of MFH occurring in the maxilla with a review of literature about the clinical behavior and treatment of this lesion.

A CASE REPORT

A 30-year old woman was referred by general dental practitioner with a four month history of swelling and discomfort of right posterior palatal area. Four months earlier she punctured the palatal swelling with a needle by herself and then the ulcer-like wound developed and didn't heal.

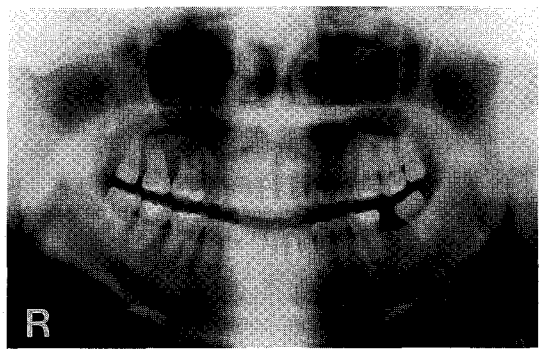

Fig. 1. Panoramic Radiograph. It shows the periapical osteolytic lesion at the right maxillary area (#13-18) with root resorption of the first and second molar teeth.

She delivered the second child just 3 days before the visit to the Department of Oral & Maxillofacial Surgery of Chosun University Dental Hospital. Examination revealed the presence of a tender ulcerative lesion on the right posterior palate. The upper second premolar tooth and the molar teeth were mobile. The roentgenograms demonstrated the periapical osteolytic lesion at the right posterior maxillary area (#13-18) with root resorption of the first and second molar teeth (#16, #17)(Fig. 1). The two molar teeth showed floating appearance. The border of the lesion was not demarcated and diffuse and the posterior wall of right maxillary sinus showed discontinuity. The elevation of the right maxillary sinus floor was suspected due to the lesion.

CT scan showed the relatively well defined and slightly lobulated homogenous mass with bone destruction in the right maxilla and the right alveolar bone (Fig. 2). The mass extended into infratemporal fossa, roof portion of oropharynx, palatine bone, inferior nasal concha and nasal cavity. The mass invaded the inferolateral and mediolateral wall of the right maxillary sinus, showing protruded mass from maxillary cavity. There was no enlarged cervical lymph node. The molars and premolars appeared floating. Radiographs of the chest showed no abnormality.

Before the operation, incisional biopsy was done via intraoral approach and it confirmed MFH (Fig.

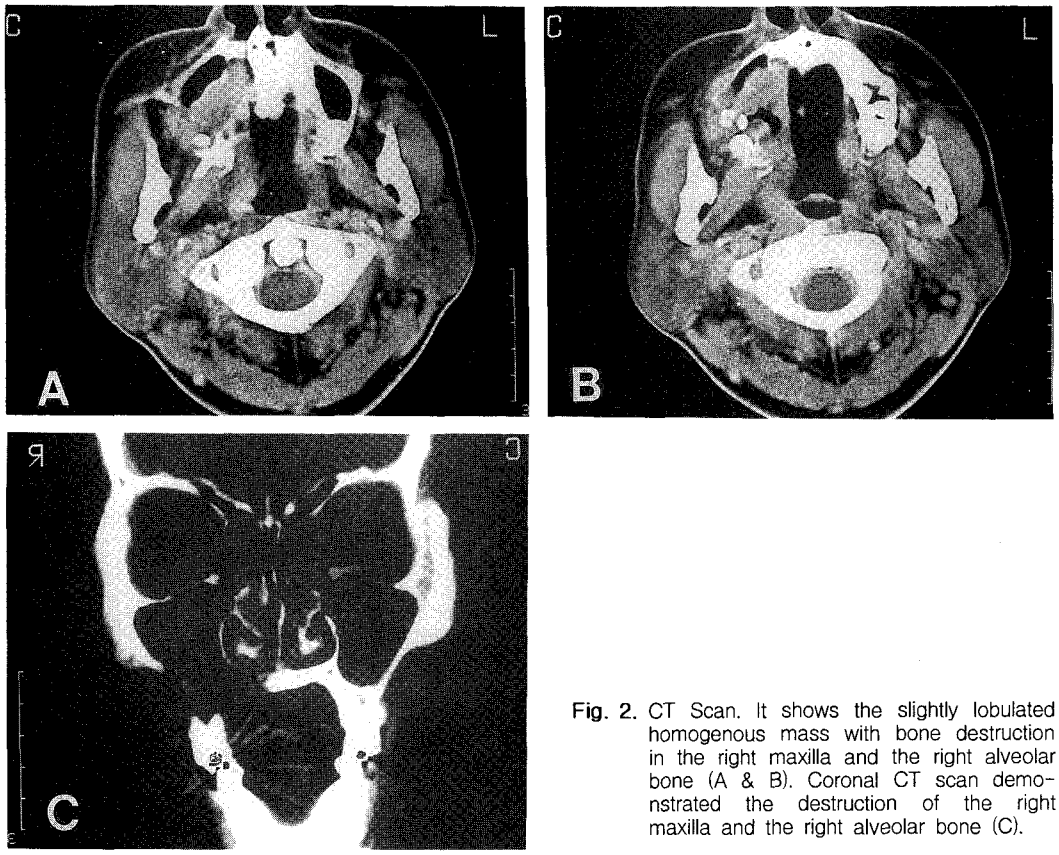

Fig. 2. CT Scan. It shows the slightly lobulated homogenous mass with bone destruction in the right maxilla and the right alveolar bone (A & B). Coronal CT scan demonstrated the destruction of the right maxilla and the right alveolar bone (C).

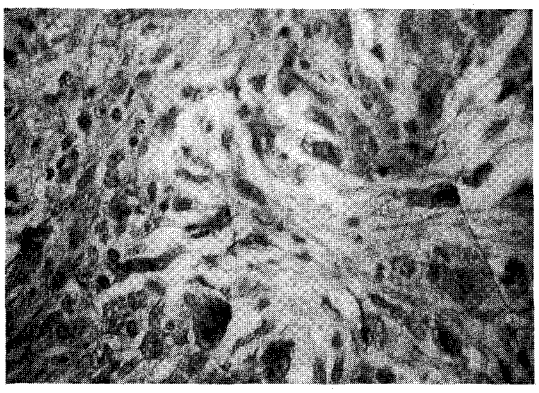

Fig. 3. Histologic Section of Malignant Fibrous Histiocytoma of the Maxilla.

3). At the time of surgery, the lesion was approached using Weber-Fergusson incision and a maxillectomy and skin graft was performed on Sep. 14, 1994. At the frozen biopsy tumor cells were seen in the external pterygoid muscle, so the surgeon excised more widely. At 4 weeks after operation she had the difficulty in opening of mouth. The mouth opening was 14 mm, and 24 mm after mouth opening exercise. A course of postoperative RT was given using Co-60 teletherapy unit (Picker Company) to the previous tumor mass area with a 2 cm margin around. Radiation therapy started in 6 weeks after operation. We used the 45° pair-wedged right lateral and anterior fields with field size of 10 × 11 cm (right lateral) and 9 × 12 cm (anterior). After the dose of 50.4 Gy parallel opposing lateral fields (8 × 6 cm) were used with weighing for the right lateral. The patient was treated with 1.8 Gy/fraction to a total dose of 59.4 Gy/ 33 fractions in 46 days.

At 5 months after operation or 2.5 months after

irradiation she couldn't eat well due to trismus (1 mm). It was treated by mouth opening exercise with mouth gag under general anesthesia. After that limited mouth opening improved and the maximum interincisal opening is 16 mm. The patient remains free of disease, 10 months post-operatively.

DISCUSSION

MFH of the maxilla is a rare malignant bone tumor. Review of 16 cases of MFH occurring in the maxilla revealed the typical presentation of maxillary sinus malignancy, such as facial swelling, pain, and loose teeth as initial presentations[9]. Block et al. reported local recurrence or distant metastases in 55% of cases of maxillary MFH[9]. Metastases in the head and neck region are found principally in the regional lymph nodes and lungs.

Forty-one MFHs of the jaws and oral soft tissues were reviewed by Happonen et al.[10]. This excludes those arising in the paranasal sinuses. Twenty-one lesions were located in the mandible and 12 in the maxilla. Three tumors involved oral soft tissues only and the exact locations of five lesions were not given. The age of the patients ranged from 11 to 81 years (mean, 42.7 years). Fourteen patients had been treated by radical surgery and 10 by a combination of surgery and radiation therapy. Adjuvant chemotherapy had been used on the treatment of nine tumors. In addition, two inoperable patients received palliative radiation only. The method of treatment was not given in 15 cases. Follow-up information was available from 26 patients, with a mean follow-up time of 27 months (range, 4 months to 8 years). Fourteen recurrences were noticed within 1 to 18 months after the primary therapy (mean, 8 months). Multiple recurrences were reported in four cases and metastatic lesions in 16 patients, most commonly in the lungs. Fourteen out of the 26 patients died of the disease. Their mean survival time was 2.5 years (range, 6 months to 8 years).

Bras et al. reviewed 16 patients of MFH involving the oral soft tissues[11]. The 2-, 3-, and 5-year survival rates were 35%, 30%, and 20%, respectively. Of the patients who died, the period of survival from time of diagnosis ranged from 1 month to 5 years with an average survival time of 16 months and a median survival time of 8.5 months. The survival rate of patients with MFH of the oral soft tissues was worse than that of patients with the deeply situated tumors, described in large series of soft tissue malignant fibrous histiocytomas. The high incidence of regional lymph node involvement was striking. The treatment of choice was early and radical surgery, including neck dissection when the tumor was situated in the oral regions. The response to chemotherapy, except for the response of patients with the inflammatory type of MFH, and radiotherapy was usually poor.

Adbul-Karim et al. reported a clinicopathologic study of 11 cases; MFH of jaws[12]. The behavior of MFH was more aggressive than that of fibrosarcoma and osteosarcoma in this site. Recurrence was common, and MFH often metastasized early in the course of the disease, most commonly to the lungs. The prognosis appeared to be poor. The management of MFH in the jaws required early radical surgery and a close follow-up. The value of adjuvant radiotherapy and chemotherapy was still questionable.

MFH of the maxilla is best treated surgically but radical neck dissection does not appear to be indicated unless there is clinical evidence of lymph node metastases[13]. The extension of maxillary MFH, along with the anatomical complexity of the retromaxillary region, make surgical access for excision difficult. Total maxillectomy via an anterior or modified Weber-Fergusson approach gives poor posterior exposure so that separation of the maxilla from the pterygoid plates is usually performed blindly, and tumor extending posterior to the maxillary sinus might remain. Further removal of the pterygoid plates and musculature is difficult after removal of the maxilla.

Although the use of radiotherapy for head and neck MFH has not been studied for a series of

cases, individual cases of regression or histological change have been reported. Hayter et al.[4] reported a case of maxillary MFH which was considered inoperable for such a large invasive tumor with probable involvement of the base of the skull and treated with radical radiotherapy, 55 Gy in 20 treatments over 28 days to the right maxilla with a 2 cm margin all the way round. When examined histologically after 13 months there was no remaining local tumor. In fact, the absence of residual tumor within the antrum at post-mortem provides evidence of the effectiveness of this mode of treatment. Other authors including Feldman, et al.[8], Blitzer et al.[7], and Soule and Enriquez report numbers of cases who received irradiation without benefit. Blitzer et al. reported that radiation therapy seems to have very limited efficacy with these tumors and should be reserved for patients who are very poor surgical risks, or who have inoperable recurrences or distant metastases. Radiation therapy has often been used adjunctively, and sometimes primarily, but dosage and fractionation, method of administration, and tumor response are not well documented.

Review of literature regarding radiation therapy in the treatment of MFH, authors analyzed of 17 patients with localized MFH who received radiation therapy[14]. Of 5 patients who were treated by irradiation alone, 2 were locally controlled: a literature survey revealed that local control was obtained in only 2 of 16 patients treated by irradiation alone. Of the 12 patients treated postoperatively, 9 were controlled locally, whereas 12 of 14 patients collected from the literature were controlled with postoperative irradiation. Local recurrence following resection is reduced from approximately 50% to 20% by post-operative irradiation. Seven of 17 patients (41%) developed distant metastases: six patients have died, and the remainders of 11 patients have been followed for 12 to 35 months (median, 25 months). Radiation therapy can eradicate small tumor masses, and given postoperatively, appears to lower risk of recurrence.

Response to combination chemotherapy in 33% of 23 patients with recurrent or metastatic MFH has been reported by Leite et al.[15]. The inflammatory type of MFH seems more sensitive to chemotherapy[11]. Chemotherapy also resulted in complete remission in five cases of skeletal MFH reported by den Heeten et al.[16]. Two first underwent a primary amputation, whereas the other three received primary chemotherapy with histologic evaluation of the effect. These patients showed a complete remission. The five patients completed chemotherapy were all still alive, without indications of metastasis or local recurrence. Although the number of cases is small, a 25- to 58- months (mean 45) survival, in 5 patients treated either with chemotherapy alone or chemotherapy and surgery, is surprisingly good in view of previous experience with this tumor. In some of these patients, the authors were able to document an absence of any viable tumor following chemotherapy with the aid of high-dose methotrexate and citrovorum factor rescue[16]. Such reports encourage the future trial of adjuvant chemotherapy in the treatment of maxillary MFH.

In this case the patient tolerated irradiation well except mucositis. The mucositis improved with conservative and symptomatic treatment. So it seems to be safe and effective to deliver postoperative irradiation because the patients are well tolerable to RT and there are the reports of benefits from postoperative irradiation. In cases of recurrences or distant metastases it seems to be possible to give combination chemotherapy.

It appears to be necessary to report uncommon tumors to provide relevant information for future study and management.

REFERENCES

1. **Fagundes HM, Lai PP, Dehner LP, et al.** Postoperative radiotherapy for malignant fibrous histiocytoma. Int J Radiat Oncol Biol Phys 1992; 23: 615-619
2. **Alvegard TA, Berg NO.** Histopathology peer review of high grade soft tissue sarcoma: the

Scandinavian Sarcoma Group Experience. J Clin Oncol 1989; 12(7):1845-1851

3. **Besly W, Wiesenfeld D, Kleid S, Allan P, Poker I.** Malignant fibrous histioicytoma of the maxilla-- a report of two cases. Brit J Oral Maxillofac Surg 1993; 31:45-48
4. **Hayter JP, Williams DM, Cannell H, Hope-Stone H.** Malignant fibrous histiocytoma of the maxilla: Case report and review of the literature. J Maxillofac Surg 1985; 13:167-171
5. **Pezzi CM, Rawlings MS, Esgro JJ, Pollock RE, Romsdahl MM.** Prognostic factors in 227 patients with malignant fibrous histiocytoma. Cancer 1992; 69:2098-2103
6. **Weiss SW, Enzinger FM.** Malignant fibrous histiocytoma: an analysis of 200 cases. Cancer 1978; 41:2250-2266
7. **Blitzer A, Lawson W, Biller HF.** Malignant fibrous histiocytoma of the head and neck. Laryngoscope 1977; 87:1479-1499
8. **Feldman F, Norman D.** Intra- and extraosseous malignant histiocytoma (Malignant Fibrous Xanthoma). Radiology 1972; 104:497-508
9. **Block MS, Cade JE, Rodriguez FH.** Malignant fibrous histiocytoma in the maxilla: Review of literature and report of a case. J Oral Maxillofac Surg 1986; 44:404-412
10. **Happonen RP, Ekfors T, Suonpaa J, Forssell K.** Malignant fibrous histiocytoma of the jaws: Report of two cases. J Oral Maxillofac Surg 1988; 46:690-693
11. **Bras J, Batsakis JG, Luna MA.** Malignant fibrous histiocytoma of the oral soft tissues. Oral Surg Oral Med Oral Pathol 1987; 64:57-67
12. **Adbul-Karim FW, Ayala AG, Chawla SP, Jing BS, Goepfert H.** Malignant fibrous histiocytoma of jaws: A clinicopathologic study of 11 cases. Cancer 1985; 56:1590-1596
13. **Ogura JH, Toomey JM, Setzen M, Sobol S.** Malignant fibrous histiocytoma of the head and neck. Laryngoscope 1980; 90:1429-1440
14. **Reagan MT, Clowry LJ, Cox JD, Rangala N.** Radiation therapy in the treatment of malignant fibrous histiocytoma. Int J Radiat Oncol Biol Phys 1981; 7:311-315
15. **Leite C, Goodwin JW, Sinkovics JG, Baker LH, Benjamen R.** Chemotherapy of malignant fibrous histiocytoma: A southwest Oncology Group Report. Cancer 1977; 40:2010-2014
16. **Heeten GJ, Koops HS, Kamps WA, Oosterhuis JW, Sleijfer DT, Oldhoff J.** Treatment of malignant fibrous histiocytoma of bone: A plea for primary chemotherapy. Cancer 1985; 56:37-40

국문초록 =

상악골의 악성 섬유성 조직구종

- 증 례 보 고 -

조선대학교 의과대학 치료방사선과[1], 조선대학교 치과대학 구강외과[2]

오 윤 경[1] · 여 환 호[2]

악성 섬유성 조직구종은 상악골부위에서는 매우 드물게 발생하는 악성 종양으로서, 두경부에 발생되는 예가 전체 악성 섬유성 조직구종의 7% 이며 이들 7%중 12%가 상악골에 발생한다고 한다.

상악골의 악성 섬유성 조직구종은 국소재발이나 원격전이가 55%의 환자들에서 발생하며 평균 생존기간은 상악골, 하악골 및 구강연부조직의 악성 섬유성 조직구종에 관한 연구에서 30개월로 보고되었다.

상악골의 악성 섬유성 조직구종은 일차적으로 근치적 수술요법이 주 치료방법이다. 방사선치료에 관한 보고들은 대상환자들이 적어서 지금까지 체계적으로 보고되지 않았고, 증례보고에 의한 종양의 퇴행 또는 조직학적 변화가 발표된 바 있으며 일부 저자들은 수술후 방사선치료가 도움이 되지 않으므로 재발성 또는 수술 불가능한 경우에 시행하기를 권하기도 한다. 재발성 또는 전이성 악성 섬유성 조직구종 환자들에서 항암화학요법으로 33%의 관해율을 보였다는 보고가 있다.

저자들은 상악골의 악성 섬유성 조직구종 1예를 경험하였기에 문헌고찰과 함께 보고하는 바이다.