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Fibrosarcoma of the Jaws

A Study of 7 Cases

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Summary

Seven cases of fibrosarcoma of the jaws are presented. Five cases were in the mandible and 2 involved the maxilla. There were 3 males and 4 females and ages varied from 11 to 59 years. Four patients were in their third decade. All patients died of their tumour. Reason for death was local extension (2 patients); metastatic spread (2 patients); both local extension and metastatic spread (2 patients); and post-operative complications (1 patient). Factors causing this unfavourable clinical course are discussed. Chemotherapy and irradiation were of no therapeutic benefit. Some notes on histogenesis are made with respect to an odontogenic origin of jaw fibrosarcoma. This possibility exists but contributory evidence is equivocal unless a secondary fibrosarcoma has arisen in a benign odontogenic tumour.

Key-Words

Malignant bone tumour – Fibrosarcoma – Histiocytic fibrosarcoma – Malignant jaw tumour

Introduction

Fibrosarcoma of bone is a tumour entity that has to be distinguished from other malignant bone tumours. It is a neoplasm which produces collagen fibres but no new bone or cartilage formation, in which case a diagnosis of osteosarcoma or chondrosarcoma must be made.

The tumour is considered to arise from the medullary connective tissue of the skeleton and must not be confused with periosteal fibrosarcoma which has its origin in soft tissues adjacent to the bone, as this latter lesion has a better prognosis (*Huvos* and *Higinbotham*, 1975). If the tumour arises in an otherwise normal bone, it is called a primary fibrosarcoma of bone whereas in the event of a pre-existing bone lesion it is referred to as a secondary fibrosarcoma. Predisposing lesions include fibrous dysplasia, Paget's disease or giant-cell tumour; also fibrosarcoma is seemingly induced by chronic osteomyelitis or irradiation. In addition, ameloblastic fibroma and myxoma have been documented as primary jaw tumours that may evolve into fibrosarcoma (*van Blarcom* et al., 1971; *Reichart* and *Zobl*, 1978).

In recent years, histiocytic fibrosarcoma has been distinguished from fibrosarcoma. This former designation is employed for lesions that show, in addition to the malignant fibroblasts, a cell population that is characterized by histiocytic features such as a foamy cytoplasm, giant cell formation and folded or grooved cell nuclei (*Spanier* et al., 1975). This histomorphology is considered to be due to a dual differentiation of the tumour cells in a fibroblastic as well as a histiocytic direction. The validity of distinguishing between fibrosarcoma and histiocytic fibrosarcoma of bone has, however, been considered questionable on histological grounds (*Dahlin* et al., 1977) and has been shown to have no clinical significance (*Freyschmidt* et al., 1981). Therefore we will discuss both entities together under the all-encompassing term fibrosarcoma.

Fibrosarcoma of bone is an uncommon disease as evidenced by a relative incidence that varies from 3.5 % (*Larsson* et al., 1976) to 12 % (*McKenna* et al., 1966) of all primary malignant bone tumours. The long bones are most often involved; figures for occurrence in the jaw bones vary from 11 % (*Taconis*, 1982) to 14 % (*Jeffree* and *Price*, 1976). Reports that are exclusively devoted to jaw fibrosarcomas are exceedingly scarce. Besides reports of single cases there is only one paper that reports an original series of 13 cases (*van Blarcom* et al., 1971) whereas *Eversole* et al. (1973) compiled a series by reviewing the pertinent literature.

Because of the paucity of data on fibrosarcoma of the jaw, we feel justified in reporting our experience with this tumour entity.

Material and Methods

The files of the Department of Oral Pathology at the Utrecht University Dental Institute were searched for cases coded as fibrosarcoma of the jaw. Over a period of 30 years (1952–1982) 14 cases were collected. Four cases were excluded from the study as they represented fibrosarcomas of the parapharyngeal soft tissues with secondary involvement of the mandible; 3 cases were inadequately documented and these were not included because proof of intraosseous origin was lacking. So there remained 7 cases that were fully documented histologically as well as radiologically and with complete follow-up data. These cases are summarized in Table 1. Cases 6 and 7 have also been the subject of previous papers (*Slootweg* and *Müller*, 1977, 1983) but are included here for the sake of completeness. All cases originated in previously normal bone; so none of them could be classified as secondary fibrosarcoma. During the above-mentioned period, a total of approximately 10 000 specimens had been submitted to the Oral Pathology Department, of these osteosarcoma was diagnosed in only 20 cases.

Clinical Features

The ages of the 7 patients at the time of the original diagnosis ranged from 11 to 59 years; 4 patients were in the third decade. Pain and swelling were the most frequent complaints, each occurring in 3 patients. Loosening of teeth, trismus, a pathological fracture or paraesthesia of the lower lip were the presenting symptoms in the other 4 patients. The interval after onset of symptoms until diagnosis and therapy varied from a few weeks to 6 months.

Radiographic Features

The tumours presented as radiolucent lesions that, in all but one cases, (case 2) exhibited poorly defined margins. Loss

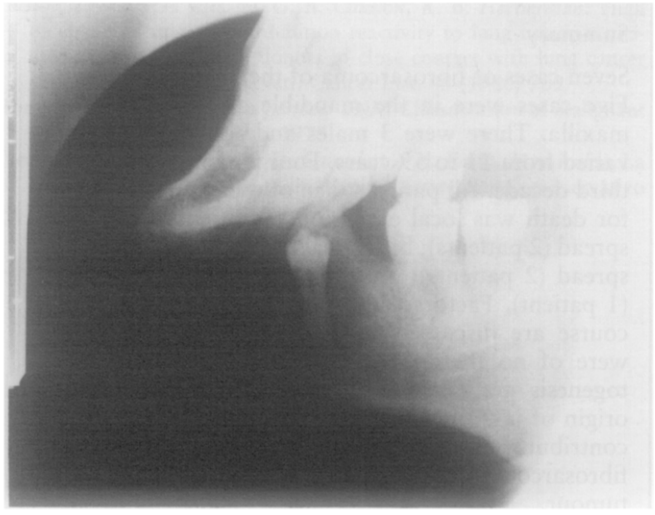

Fig. 1 Case 2. Radiograph shows radiolucent lesion surrounding the crown and the coronal half of the root of the impacted 35.

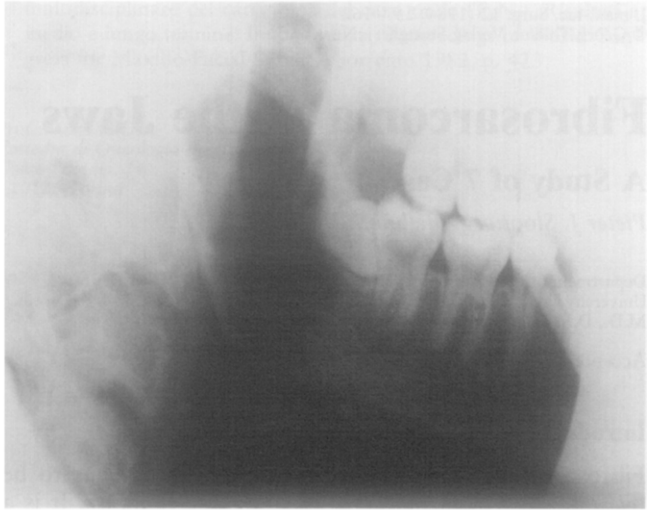

Fig. 2 Case 3. Radiograph shows radiolucent lesion within the mandibular ramus.

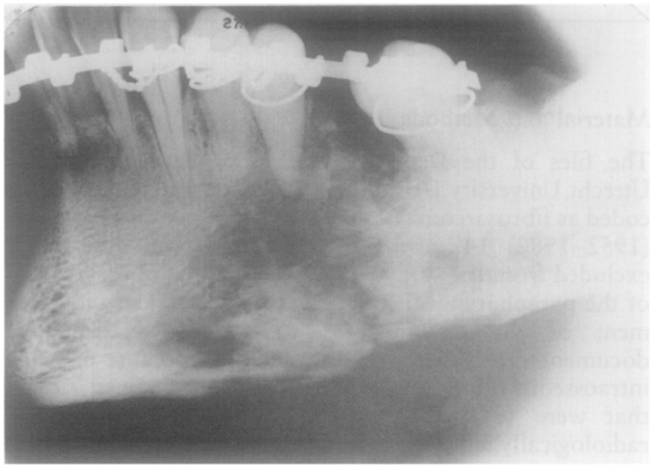

Fig. 3 Case 4. Radiograph of surgical specimen clearly displays moth-eaten destruction of mandibular body causing a pathological fracture.

of alveolar bone suggested a "hanging in the air" appearance of the involved teeth, in 2 cases (cases 5 and 7). A moth-eaten bone destruction as evidenced by multiple small and irregular radiolucencies was seen in case 4 in which there was also a pathological fracture. In case 2, a rather well circumscribed radiolucency surrounded the crown and coronal half of the root of an impacted premolar tooth. Presumptive diagnosis in this case was follicular cyst or benign odontogenic tumour. The various radiographic appearances are shown in Figs. 1–4.

Gross Morphology

Two surgical specimens (from cases 4 and 5) were available for macroscopic evaluation. Both tumours were soft white masses with a haemorrhagic and necrotic centre (Figs. 5 and 6). Infiltration of bone was obvious and perforation of the cortical plates had resulted in poorly demarcated extension into the adjacent soft tissues. One other specimen (from case 2) was reported to be composed of a firm-elastic

Table 1 Clinical data of reported patients

Case no.	Age*	Sex	Location	Therapy	Clinical course**	Follow-up**
1.	11	F	right mandibular ramus	irradiation	local recurrence after 1 yr.	died after 2 yrs.
2.	50	M	left mandibular body	resection	lung metastasis after 1 yr.	died after 2½ yrs.
3.	24	F	right mandibular ramus	resection	local recurrence after 4 yrs.	died after 4½ yrs.
4.	30	F	left mandibular body	resection, irradiation, chemotherapy	local recurrence and metastasis to lung, brain, skin and skeleton after ½ yr.	died after 1 yr.
5.	28	F	right mandibular body	resection, irradiation, chemotherapy	skeletal metastasis after ½ yr.	died after ½ yr.
6.	59	M	left maxilla and cervical lymph node metastasis at time of admission	resection, neck dissection and irradiation	brain abcess as postoperative complication	died after ½ yr.
7.	26	M	left maxilla	resection	3, 4, and 5 yrs. later recurrence treated with re-excision, irradiation, chemotherapy	died after 5 ½ yr. with local and metastatic disease (brain, lung, pleura)

*Age in years at time of diagnosis
**Time interval after initial diagnosis and treatment

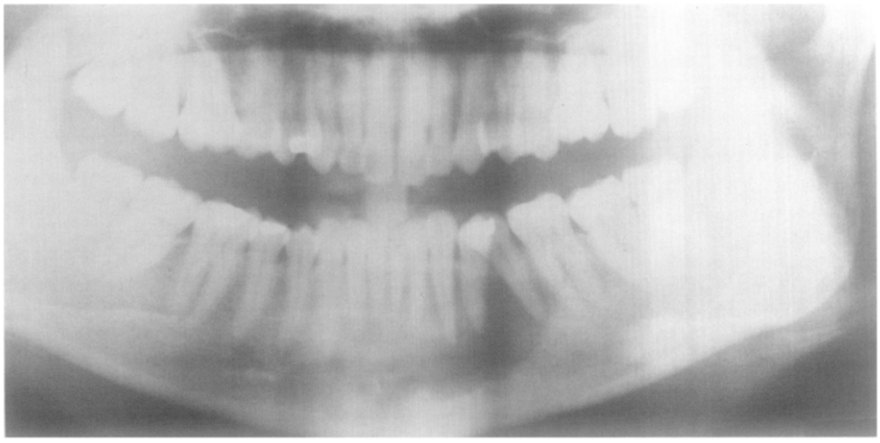

Fig. 4 Case 5. Radiograph shows loss of bone in the area 42–35 suggesting a "hanging in the air" appearance of the involved teeth.

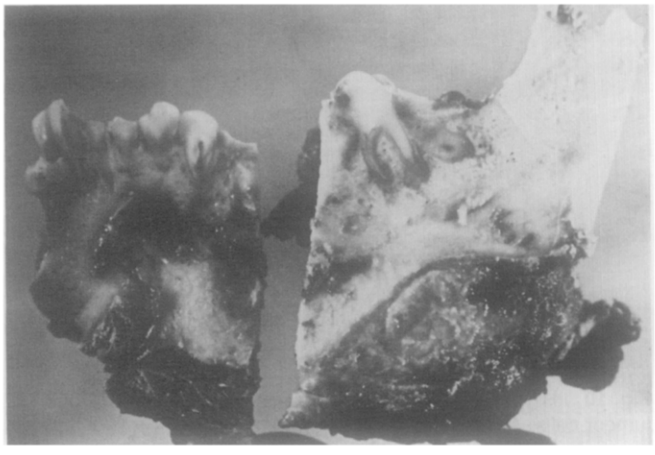

Fig. 5 Hemisectioned surgical specimen from case 4 shows cortical perforation and spread of the tumour in the adjacent soft tissues.

Fig. 6 Hemisectioned surgical specimen from case 5 reveals extensive destruction of the mandibular body with perforation of lingual as well as buccal cortical plates.

white tumour mass that filled a cavity with an irregular outline in the mandible. As the specimen was not available, we were not able to confirm this by personal examination. Macroscopic data on the other cases was not available.

Histopathological Features

All tumours are composed of pleomorphic spindle cells with varying degrees of differentiation and the capacity to produce collagen fibres. The "herring-bone" pattern of interlacing bundles that are arranged perpendicular to each other is sometimes very pronounced (Fig. 7). In other lesions the tumour fascicles exhibit a peculiar whorling that is recognized as a storiform or "cart wheel" pattern (Fig. 8). The tumours can be divided into well and poorly differentiated types. In the former (cases 1 and 7) the individual tumour cells are clearly fusiform with oval to elongated nuclei and they are separated by bundles of collagen fibres. Mitotic figures are found but these are not very frequent (Figs. 9 and 10). In the less well differentiated tumours (cases 2–6), the tumour cells exhibit large, irregularly outlined and vesicular nuclei. The cytoplasm may be extremely scanty, only forming a small rim around the nucleus. The intercellular substance is greatly reduced and the tumour cells are closely packed. Mitotic figures are present in large numbers (Fig. 11).

In one tumour (case 3), there are large myxoid areas that contain vesicular and swollen cells with a cytoplasm drawn out in multiple delicate processes (Figs. 12 and 13). The combination of this morphology with the classical fibrosarcoma pattern elsewhere in the lesion justifies the diagnosis of myxofibrosarcoma.

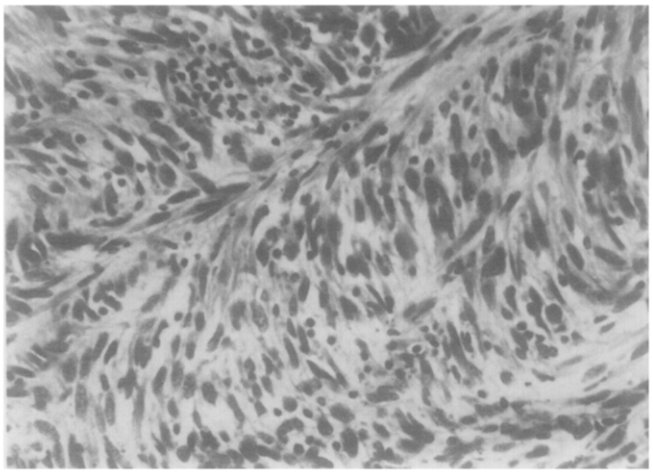

Fig. 7 Photomicrograph shows the classical "herringbone" pattern of the fibrous fascicles typical of fibrosarcoma (H & E, ×96).

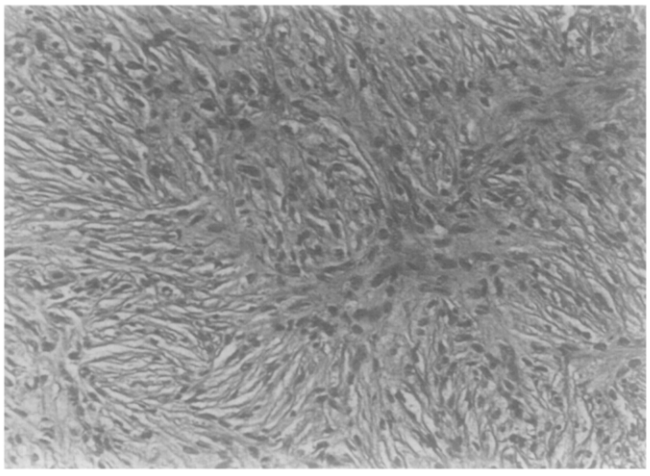

Fig. 8 Storiform or "cart-wheel" arrangement of fibrous fascicles that suggests a diagnosis of histiocytic fibrosarcoma (H & E, ×60).

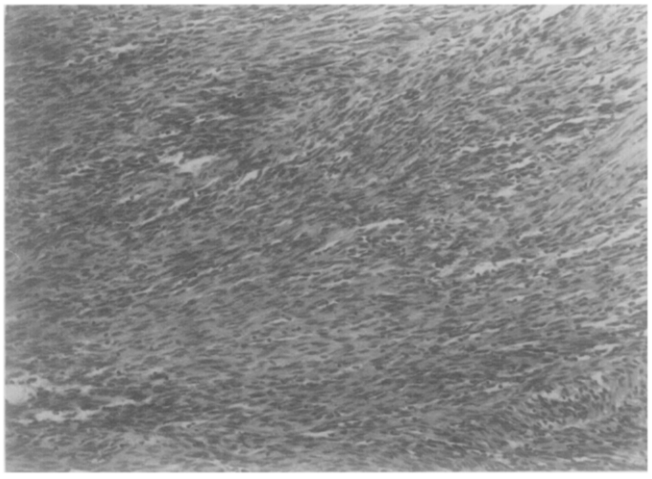

Fig. 9 Well-differentiated fibrosarcoma consisting of relatively cell-poor fibrous bands (H & E, ×36).

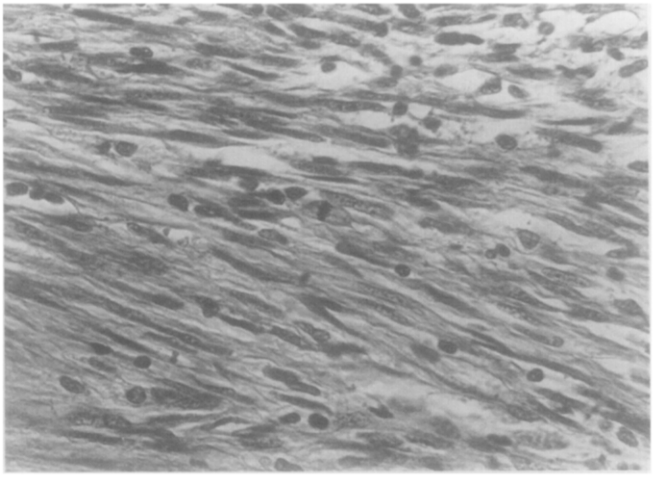

Fig. 10 High-power view of Fig. 9 shows the uniform nuclei of the tumour cells. A mitotic figure is also present (H & E, ×150).

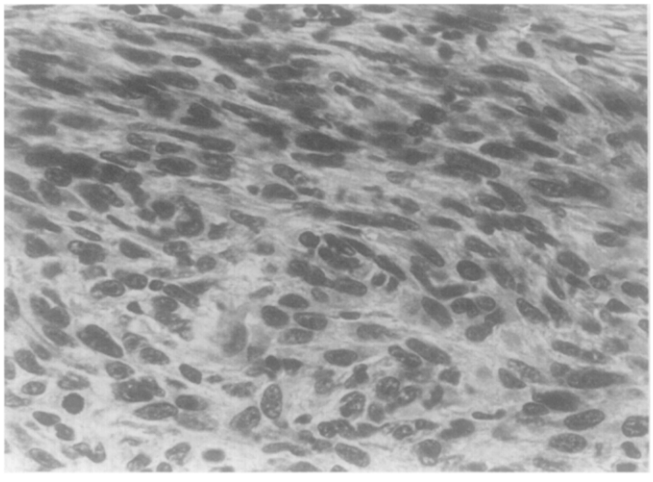

Fig. 11 High-power view of a poorly differentiated fibrosarcoma. Nuclear pleomorphism is a pronounced feature (H & E, × 150).

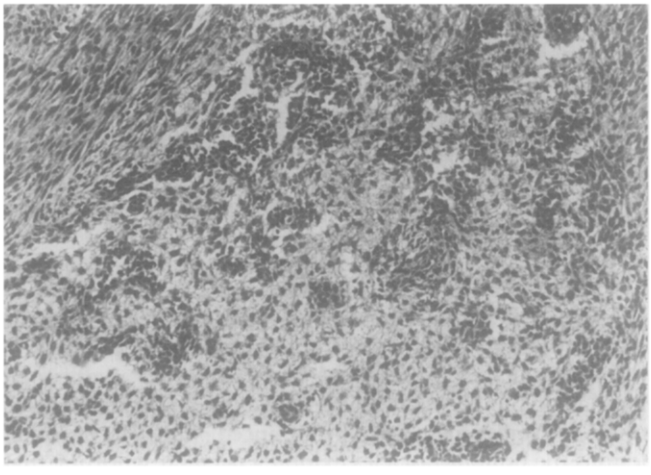

Fig. 12 Low-power view shows myxomatous tumour areas with adjacent fibrous parts. Diagnosis was myxofibrosarcoma (H & E, ×150).

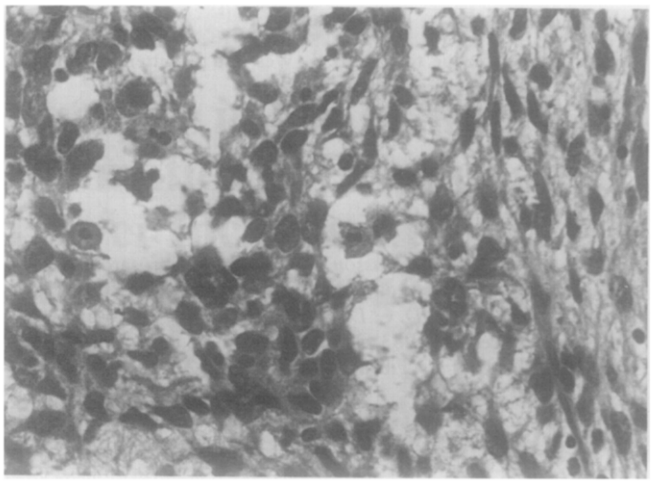

Fig. 13 High-power view of Fig. 12 shows pleomorphic cells with tapered cytoplasmic extensions in a myxomatous background (H & E, ×150).

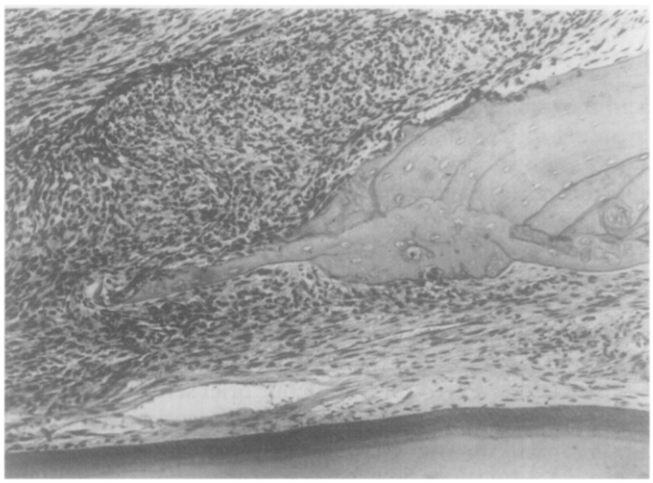

Fig. 14 Photomicrograph shows resorption of pre-existing alveolar bone and infiltration of the tumour into the periodontal ligament space. The root dentine (lower border) does not show any resorption (H & E, ×36).

Infiltration between osseous trabeculae and penetration of the cortex are obvious features. The remnants of the original bone exhibit signs of osteoclastic resorption, and cortical perforation is followed by a subperiosteal centrifugal growth pattern (Fig. 14). The macroscopic impression of central necrosis and haemorrhage is substantiated by the microscopic findings.

A storiform arrangement of the fibrous fascicles is observed in three cases (2, 6, 7). This would be considered evidence for a diagnosis of histiocytic fibrosarcoma but we consider them as fibrosarcomas for reasons as outlined previously.

Treatment and Follow-up

All but one case (case 1) had been treated by block resection of the affected jaw segment. In case 1, the tumour, although originating in the mandible had eroded the skull base and was considered to be inaccessible to surgery. This patient received irradiation as single modality treatment. Irradiation and chemotherapy were employed as adjuvant therapy in some other patients. In spite of attempts to cure, all patients died of their tumour – the longest survival time being 5½ years. Reasons for death were: – local persistence of disease (2 patients), both local and metastatic disease (2 patients), metastatic disease (2 patients), and post-operative complications (1 patient). Metastasis to the lungs developed in 3 patients, to distant bones in 3 patients, to the brain in 2 patients and cutaneous metastasis was observed in 1 patient. One patient showed metastatic deposits in regional lymph nodes of the neck (case 6).

Discussion and Conclusions

Differentiation between fibrosarcoma of bone and soft-tissue fibrosarcoma has prognostic significance. For patients with soft-tissue fibrosarcoma the five-year survival rate approximates 60 % (*Pritchard* et al., 1977) as opposed to a five-year survival rate of fibrosarcoma of bone that varies from 4.2 % (*Larsson* et al., 1976) to 31.7 % (*McKenna* et al., 1966). Fibrosarcomas that arise in the periosteum – the so-called periosteal fibrosarcoma – exhibit the better prognosis in soft-tissue fibrosarcoma, and so, differentiation between fibrosarcoma of bone arising in the bone marrow and periosteal fibrosarcoma is a rewarding exercise (*Huvos* and *Higinbotham*, 1975).

It is a firmly established fact that osteosarcomas of the jaw differ in clinical course from those arising in the long bones by their exhibition of a far lower rate of metastasis (*Clark* et al., 1983). A similar difference in clinical behaviour between fibrosarcoma of jaw bone and fibrosarcoma of the long bones has been reported in the literature as a consistent finding. *Taconis* (1982) makes mention of a 71 % five-year survival of patients with jaw fibrosarcoma as opposed to a 38 % five-year survival rate for those with long bone fibrosarcoma; a difference that he also found for a ten-year survival period: 32 % for the long bone group in comparison with 50 % for the cases with jaw involvement. A similar better prognosis for jaw fibrosarcoma has been reported by *Jeffree* and *Price* (1976) who found that 3 out of 4 patients with fibrosarcoma of the jaw survived over 3 years and 2 for over 10 years. In the Mayo Clinic series of bone fibrosarcomas, mandibular fibrosarcoma exhibited a five-year survival rate of 40 % which is a more favourable outcome than the 28.7 % found in the whole series (*Dahlin* and *Ivins*, 1969; *van Blarcom* et al., 1971). In a series from the Memorial Sloan Kettering Cancer Center, the better prognosis for jaw fibrosarcoma only became apparent in the long term. The five-year survival rate was 27 % for both long bone and jaw cases whereas this figure decreases to 17 % after 20 years for the long bone fibrosarcomas but remains constant at 27 % for jaw fibrosarcoma throughout a 20-year-observation period (*Huvos* and *Higinbotham*, 1975).

In our own small series, the five-year survival rate for fibrosarcoma of the jaw is zero. All patients died of their tumour after a period that varied from ½ to 5 years. These poor results may be due to the following factors. Both patients with well-differentiated tumours (cases 1 and 7) presented for treatment with tumours of a size that precluded adequate surgical intervention. All authors agree that the only chance of survival for patients with fibrosarcoma of bone lies in complete surgical removal of the local

lesion (*Dahlin* and *Ivins*, 1969; *Huvos* and *Higinbotham*, 1975; *Jeffree* and *Price*, 1976). Another patient (case 6) died of post-operative infection that spread into the brain and the remaining patients all suffered from a tumour with a poorly differentiated histomorphology as evidenced by a high mitotic frequency and a pronounced cellular pleomorphism coupled with scanty production of collagen. Low grade differentiation influences survival in a negative way (*Dahlin* and *Ivins*, 1969; *Jeffree* and *Price*, 1976) and the more favourable clinical course of jaw fibrosarcoma has been ascribed to a relative predominance of well-differentiated tumours in the jaw in comparison with the rest of the skeleton (*Taconis*, 1982). On account of the above mentioned factors, a zero survival in our brief series is due to (1) a relative predominance of poorly differentiated tumours, (2) the size of the well-differentiated tumours that ruled out adequate surgical therapy, and (3) post-operative complications.

Converse reasoning makes it apparent that our series underscores the opinion that poorly differentiated tumours have a grave prognosis and that adequate surgery offers the best chance for a cure. In our hands, irradiation and chemotherapy were of no use. Metastatic lesions continued to increase in size while the patients (case 4 and 5) were on chemotherapy.

With respect to histomorphology, no significant features differing from those observed by others were encountered. One case (3) exhibited myxomatous areas apart from classical fibrosarcoma and could be designated as myxofibrosarcoma. This appearance has also been noted by *Dahlin* and *Ivins* (1969) in fibrosarcomas of the extra-gnathic skeleton and so there are no reasons to suppose any origin of this tumour from a pre-existing jaw myxoma. This is not to say that in a particular case a fibrosarcoma cannot arise from a pre-existing jaw myxoma. Such a form of secondary fibrosarcoma has been mentioned by *van Blarcom* et al. (1971).

In the past attempts have been made to distinguish between fibrosarcomas of jaw bone originating from endosteal connective tissue and fibrosarcomas derived from odontogenic mesenchyme (*Thoma* and *Goldman*, 1960). Other authors (*Gullifer*, 1936; *Reade* and *Radden*, 1966) have noted that it is difficult to document the origin of a particular jaw fibrosarcoma.

The radiological features in one of our cases (case 2) offered speculative evidence that an origin from odontogenic mesenchyme may indeed be possible as in this case the tumour was located around the crown of an impacted tooth and may have arisen in the tooth follicle. Extensive loss of alveolar bone as was noted in cases 5 and 7 could also point towards an origin from odontogenic tissue – the periodontal ligament – but the possibility of secondary infiltration by tumour arising centrally in the mandibular bone marrow cannot be excluded with certainty. These cases serve to illustrate that jaw fibrosarcomas may have an odontogenic origin but evidence for this is often equivocal. Only in the event of a secondary fibrosarcoma arising in a benign odontogenic tumour, can the odontogenic origin of a jaw fibrosarcoma be considered to be proved beyond doubt.

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