AMELOBLASTIC FIBROSARCOMA
(a case report and review of the literature)

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SUMMARY: Ameloblastic fibrosarcoma. A twenty-eight year old Turkish man with a tumor in the mandible diagnosed as "ameloblastic fibrosarcoma" is presented. Preoperative incisional biopsy was diagnosed as "ameloblastic fibroma". Histological examination of the resection speciment revealed the features of an ameloblastic fibroma in the majority of the sections. In a few sections some mesenchymal foci showed distinct cellular pleomorphism of an anaplastic sarcoma by which we considered the tumor as malignant. Relative scarcity of malignant areas gave us the idea that the process of malignant transfromation from an ameloblastic fibroma was recent.

The literature about "odontogenic sarcoma: ameloblastic fibrosarcoma and ameloblastic odontosarcoma" is reviewed.

Key words. Ameloslastic fibrosarcoma-Ameloblastic odontosarcoma-Odontogenic sarcoma.

Ameloblastic fibrosarcoma is defined as "a neoplasm with a similar structure to the ameloblastic fibroma, but in which the mesodermal component shows the features of a sarcoma" (Pindborg and Kramer 1971). Ameloblastic odontosarcoma is defined as "a very rare neoplasm, similar to the ameloblastic sarcoma, but in which limited amounts of dysplastic dentine and enamel have formed" (Pindborg and Kramer 1971).

Both tumors are classified as the subgroups of the rare tumor "odontogenic sarcoma". We have found 11 cases of ameloblastic odontosarcomas and 33 cases of ameloblastic fibrosarcomas (including the present case) with a total number of 44 cases reported in the literature (Table 1).

Case report

Our case V.B. is a 28 year-old Turkish man. He is a professional driver. His first complaint was an ill-defined pain on the right mandible in June, 1983. It was considered as a tooth-ache and a dentist extracted a right lower tooth. After the tooth extraction he described a progressive swelling on his right mandible. Eight months later he was admitted to Plastic Surgery of Istanbul Medical Fauculty in February, 1984. Radiologically, the tumor appeared as a unilocular area of radiolucency with indistinct outlines (Fig. 1). An incisional biopsy was diagnosed as: Ameloblastic fibroma. Then total excision of the lesion with partial mandible resection was performed and the histopathological diagnosis is: Ameloblastic fibrosarcoma.

The patient is alive and well 1 year and two months after operation with no recurrence. Gross findings. A partially resected mandible specimen measuring 7 × 6 × 5 cm. It mainly consisted of a tumor mass surrounding and fixed on the mandible in three sides except the upper side which had a portion of gingiva carrying three teeth with a space of lacking tooth between them. Outer surface of the tumor seemed encapsulated and smooth but on the section surface the bone was destructed. And no well-defined margination with the tumor was present. The tumor tissue had an hardly elastic consistency. The section surface was pinkish-gray with central small areas of bleeding and cystic degeneration. The largest cystic space was 0,3 cm. in diameter (Fig.2).

Histopathological findings. For histopathological study, the sections are stained with Hematoxylin and Eosin, Masson's trichrome stain, Wilder's silver stain and Periodic acid-Schiff (PAS). There are numerous islands and cords of odontogenic epithelium,

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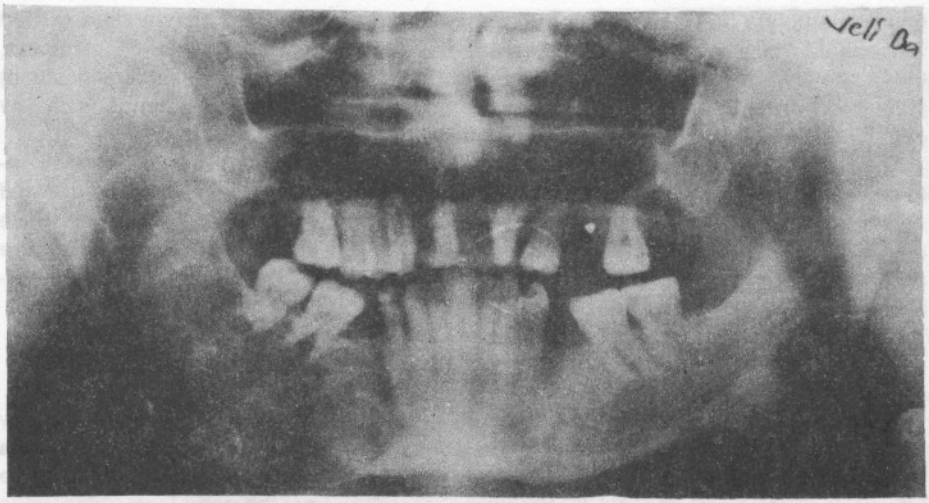

Fig.1. Radiograph showing the radiolucent lesion in the right mandible.

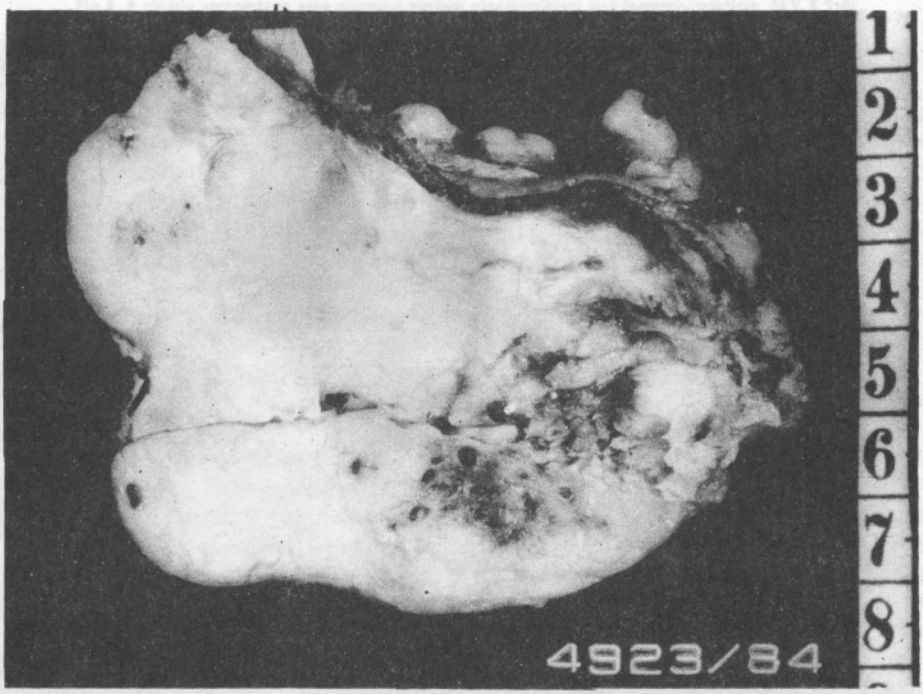

Fig.2. Cut surface of the resected specimen.

often resembling the dental lamina of early tooth development. The islands are lined with cuboidal cells intermingled with keratinized epidermoid cells As in ameloblastoma, these seets were surrounded by a basement membran (juxta-epithelial hyalinization) stainable with H.E, PAS, Masson's trichrome and reticulin staining methods. A few epithelial islands had central cystic degeneration.

The mesenchymal component mainly consists of myxomatous tissue very poor in intercellular collagenous matrixial elements. The mesenchymal cells have round or ovoid pale-staining nuclei. Some areas are hypocellular and hyalinized. Besides all these features resembling a typical ameloblastic fibroma, in a few sections there are bizarre cells with multiple or lobulated nuclei. In these foci nuclear hyperchromasia, pleomorphism and mitotic figures are present (Fig. 3,4 and 5).

DISCUSSION

Cases may arise de novo (Cases 7,9,13,16,26,36,38, present case) or they may develop by malignant transformation of their benign counterparts: Ameloblastic fibroma or ameloblastic fibroodontoma (Cases 10,11,32,33,35,37,39,43 and the majority of the others). It must be emphasized that sufficient sampling from the pathological specimen is necessary not to misdiagnose a malignant lesion since the malignancy might have begun in focal areas (Case 28,37 present case). This fact also explains the error potential of incisional biopsies (Cases 31,34, present case).

To explain the mechanism of malignant transformation some authors put forward the role of surgical traumatism for recurrent benign tumors (Kegal 1932; Dahlin and Ivins

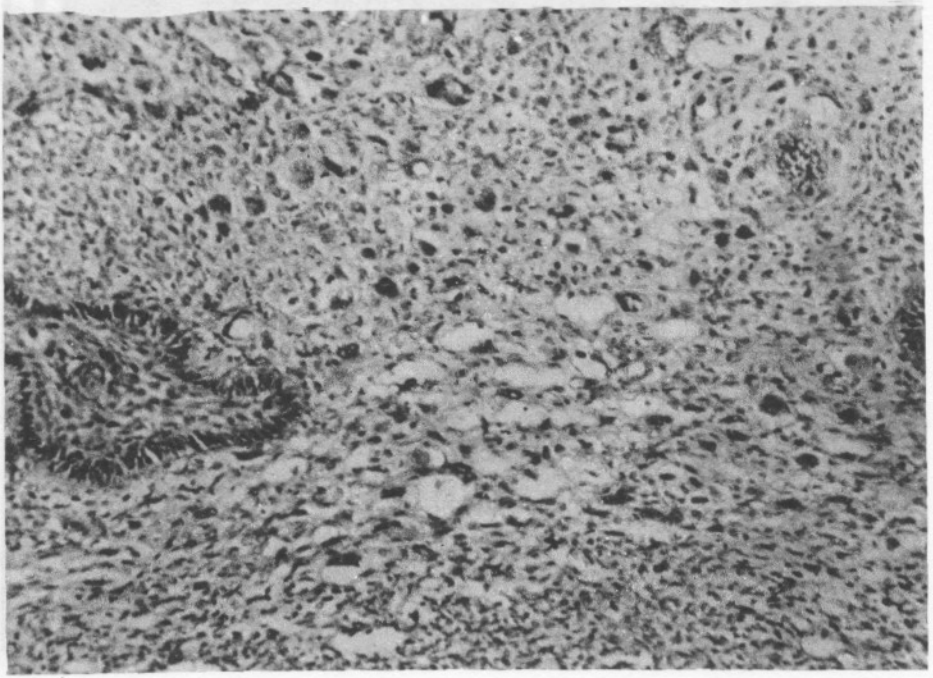

Fig.3. Microscopic appearance of the tumor showing epithelial elements and sarcomatous stroma. (H,E,X125).

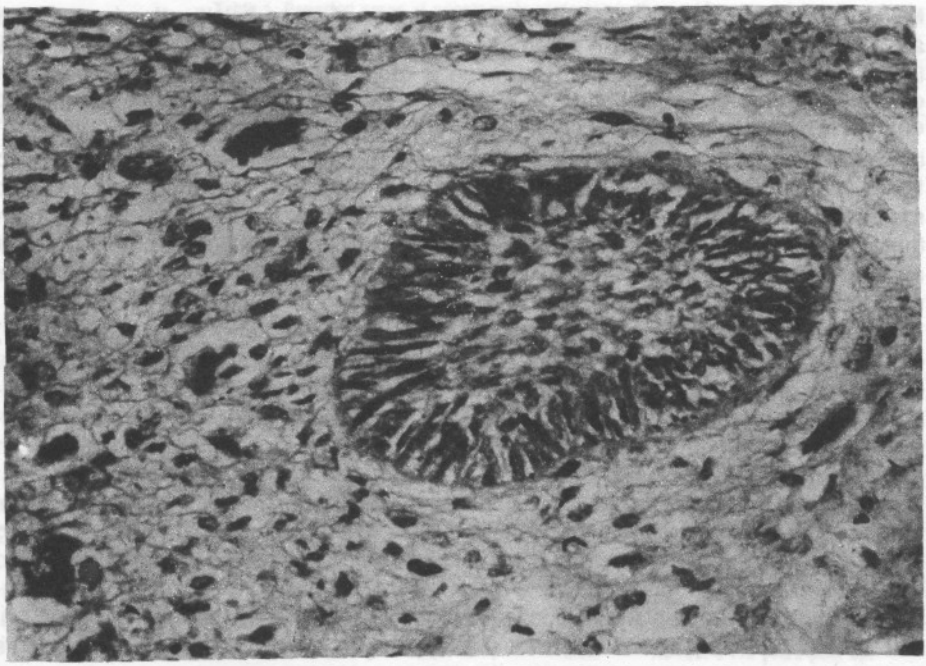

Fig.4. A clearly sarcomtous area showing nuclear pleomorphism and hyperchromatism. (H.E,X,310).

1969; Leider et al. 1972; Howell and Burkes 1977). It is also stated that the functional involution (for example numerous keratinized epidermoid cells) of the epithelial component may play a role in the sarcomatous transformation of the mesenchymal component (Chomette et al. 1983). The presence of de novo malignant cases including the present case does not support the idea of traumatic factor. We also don't agree with the idea of epithelial involution (keratinization) as a factor in malignant change. Keratinization in ameloblastoma was first mentioned in 1958 (Pindborg and Weinmann 1958). Then this property was noticed as a common feature. Now, squamous structures are considered "as an essential property of tumor cells in follicular type of ameloblastoma" (Nasu and Ishikawa 1983).

We don't have an alternative speculation about the mechanism of sarcomatous change.

Pain and swelling are he most constant findings. Ulceration, bleeding and paresthesia may be additional local findings. Pain preceeding swelling is also noted (Thoma 1951).

The histological pattern of this sarcoma is usually characteristic to allow the diagnosis. Sometimes, nuclear abnormalities and numerous atypical mitotic figures are found within a recurrent ameloblastic fibroma or fibroodontoma which undergoes a malignant change. Sometimes ameloblastic nests are intermingled with an obvious sarcomatous stroma. The amount of odontogenic epithelium may decrease or disappear in the recurrent cases (Dahlin and Ivins 1969; Peychl and Sazama 1971; Leider et al. 1972; Hatzifortiadis and Economou 1973; Reichart and Zobl 1980.) On the other hand, there are some cases showing that many ameloblastoid epithelial masses are scattered throughout the sarcomatous mesenchymal area even in the fatal stage (Pindborg 1960; Cina et al. 1962;

Mori et al. 1972; Takeda et al. 1984). The presence of some calcification is regarded as "degenerative changes" in some cases (Leider et al. 1972). Squamous changes in the epithelium are also noted (Chomette et al. 1983; Takeda at al. 1984; peresent case). According to the definiton of WHO, in the presence of dysplastic dentin and enamel formation, the tumor will be called ameloblastic odontosarcoma. Comparison between 11 cases of ameloblastic odontosarcomas (25 %) and 33 cases of ameloblastic fibrosarcomas (75 %) within the criteria of age, sex, location and prognosis gives no meaningful differences. Therefore we want to indicate our tendency to evaluate both tumors under the major heading of "odontogneic sarcoma" as do the others (Cina et al. 1962; Leider et al. 1972). Dysplastic dentin or dentin + enamel formation in the tumor may be some additional descriptive features.

Eda et al. (1976), Chomette et al. (1983), Nasu et al. (1984) and Takeda et al. (1984) examined their cases ultrastructurally. According to their common findings, the epithelial component shows a close similarity to that of ameloblastoma. In the sarcomatous component the predominant cell type closely resembles a fibroblast and evidence of varying degrees of differentiation to histiocytes, fibrohistiocytes, myofibroblasts is found in primitive mesenchymal cells.

Most of the cases are treated by radical resection of the jaw. Some less agressive surgery is noted to increase the possibility of recurrences (Cina et al. 1962; Pindborg 1960; Leider et al. 1972). Chemotherapy in case 33 with actinomycin D, Vincristine and Cytoxan is stated to produce complete response but the patient died of a second malignant tumor (malignant melanoma) in the fifth year of the treatment (Goldstein et al. 1976).

It seems paradoxical but it is true that althought the histological picture seems

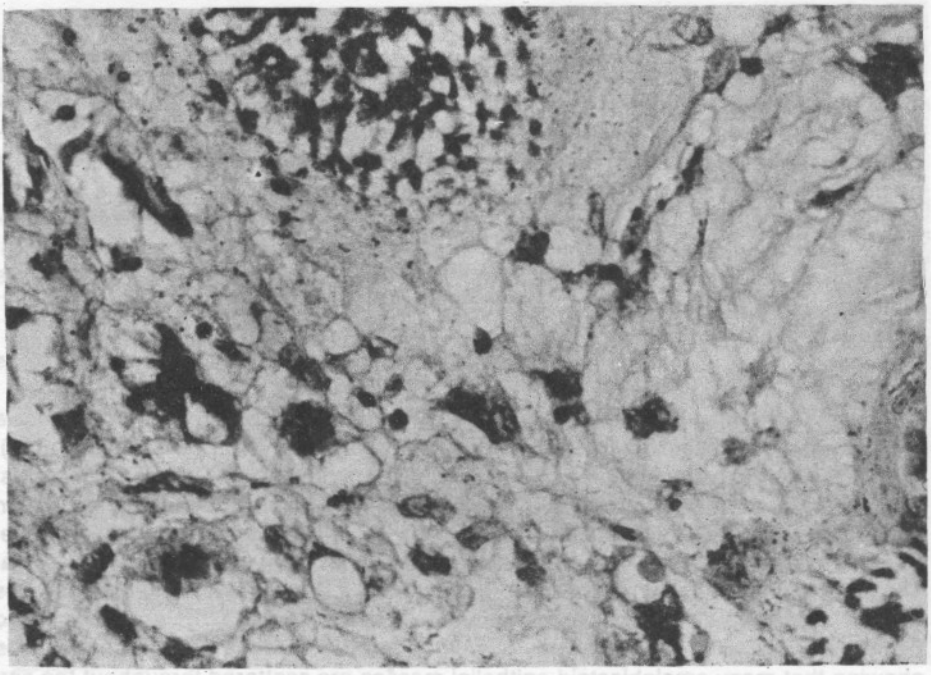

Fig.5. Bizarre stroma with atypical mitoses and distinct pleomorphic cells. (H.E,X,500).

Table 1. Reported cases of odontogenic sarcomas in the world

Case No:	Author	Year	Age (years)	Sex	Location	Follow-up
1.	Krompecher	1918	13	M	Mand.	Death at operation
2.	Papadimitriou	1928	28	F	Mand.	No rec.6/12 year post op.
3.	Kegel	1932	45	M	Max.	?
4.	Hauenstein	1937	?	M	Mand.	?
5.	Emminger	1946	52	M	Mand.	No rec. 3/12 year post op.
6.	Hertz	1952	38	F	Max.	1 rec. 12 years. post op.
7.	Thoma	1954	17	F	Mand.	No rec.7 years.post op.
x8.	Villa	1955	20	F	Mand.	No rec.4 years post op.
9.	Pindborg	1960	17	M	Max.	Death in 2^{3}/12 years with 9 recs.
10.	Cina et al.	1962	39	M	Mand.	Death in 18 years with 2 recs.
11.	Cina et al.	1962	32	F	Mand.	Death in 2^{6}/12 years with 4 recs.
12.	Muroya and Shigematsu	1962	43	M	Mand.	Death in 2^{9}/12 years with 1 recs.
13.	Cataldo et al.	1963	78	F	Mand.	?
x14.	Tahsinoğlu and Özmerzifonlu	1964	22	F	Mand.	?
x15.	Hogeman and Willmar	1966	42	M	Mand.	?
16.	Peychl and Sazama	1971	17	M	Max.	Death in 4^{6}/12 years.
17.	Leider et al.	1972	26	F	Mand.	1 rec. 16 years post op.
18.	Leider et al.	1972	43	M	Mand.	2 recs 3^{2}/12 years post op.
19.	Leider et al.	1972	9	M	Mand.	No rec. 5 years post op.
20.	Leider et al.	1972	22	F	Max.	No rec. 3^{6}/12 years post op.
21.	Leider et al.	1972	23	F	Mand.	2 recs 2^{10}/12 years post op.
22.	Leider et al.	1972	12	F	Max.	1 rec. and ?
x23.	Forman and Garrett	1972	17	M	Mand.	No rec 1^{2}/12 years post op.
(a)24.	Mori et al.	1972	3	F	Mand.	Death in 5 years with 2 recs.
25.	Mori et al.	1972	40	F	Mand.	Death in 19 years with 4 recs.
26.	Hatzifotiadis and Economou	1973	15	M	Max.	Death in 1^{9}/12 years.
(b)27.	Motegi et al.	1975	29	M	Max.	No rec. 3 years post op.
x28.	Altini and Smith	1976	27	M	Mand.	?
x(c)29.	Barbosa J.	—	36	F	Mand.	?
x(c)30.	Payen J.	—	21	F	Mand.	?
(d)31.	Eda et al.	1976	13	F	Mand.	No rec. 3 years post op.
x32.	Howell and Burkes	1977	18	F	Mand.	Death in 3^{10}/12 years with 4 recs, clinical evidence of metastasis to nerve branches.
x(3)33.	Howell and Burkes	1977	36	M	Mand.	Death in 5 years because of malignant melanoma following a chematherapy.
34.	Adekeye et al.	1978	27	M	Max.	No rec. 6/12 year post op.
35.	Reichart and Zobl	1978	16	M	Mand.	No rec. 1^{11}/12 years post op.
36.	Daramola et al.	1979	19	M	Mand.	Death early post op. because of secondary bleeding.
37.	Prein et al.	1979	24	M	Mand.	No rec. 4 years post op.
x38.	Chomette et al.	1983	9	?	Max.	No rec. 10/12 year post op.
39.	Chomette et al.	1983	38	M	Mand.	4 recs. 4 years post op.
x40.	Chomette et al.	1983	27	M	Mand.	Death in 10 years with 3 recs. and histologically proved pleuropulmonary, mediastinal lymphnode, hepatis metastases.
41.	Nasu et al.	1984	24	F	Mand.	No rec. 4 years post op.
42.	Nasu et al.	1984	29	M	Max.	No rec. 9 years post op.

43. Takeda et al.	1984	19	M	Max.	Death in 9th/12 years with 5 recs.
(f)44. Edalı et al.		28	M	Mand.	No rec. 2 years post op.

Mean age = 26.8, Median age = 24; 25M(58%), 18F(42%); 32 Mand. (73%), 12 Max. (27%).
Abreviations: M = MAle, F = Female, Mand = Mandible, Max = Maksilla, rec = recurrence, Post op = post operatively.
x = These cases show evidence of ameloblastic odontosarcoma according to WHO classification.
(a) = Also reported by Shimozato et al. (1959).
(b) = Also reported by Tagaki and Ishikawa (1972).
(c) = These cases are not published. Cited by Altini and Smith (1976).
(d) = Also reported by Eda et al. (1973).
(e) = Also reported by Goldstein et al. (1976).
(f) = Present case.

anaplastic, clinical behavior is more favorable than the conventional fibrosarcoma of head and neck (Swain et al. 1974; Huvos and Higinbotham 1975). Recurrences are noted in 15 cases (54 %) among 28 cases which have a follow-up more than 1 year. The real recurrence rate must be assumed higher than this value since the related cases might have developed recurrences after the point of their follow-up period when they were reported. Metastasis is histopathologically proved in 1 case (Case 40). In case 32, clinical evidence of metastasis to nerve branches 6,7,8 and/or brachial plexus is noted without histological examination. 14 cases are reported to have died: 2 deaths are related with operations (Case 1,36), 1 death from malignant melanoma (Case 33), 10 deaths with large local spreads and 1 death with pleuropulmonary metastases.

Related to these prognostic features is the suggestion that amelobastic fibrosarcoma is a low grade fibrosarcoma (Leider et al. 1972; Reichart and Zobl 1978) or a semi-malignant lesion (Prein et al. 1979).

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