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The radiological appearances of telangiectatic osteosarcoma

A study of 14 cases

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Abstract. Fourteen telangiectatic osteosarcomas are reported. They are rare, clinically and radiologically aggressive lesions, involving mainly the femurs of young patients, often misdiagnosed as aneurysmal bone cysts. An explanation for a characteristic early radiological appearance consisting of regular parallel striations of the shaft is suggested.

Key words: Telangiectatic osteosarcoma – Bone tumors

Telangiectatic osteosarcoma is a rare and very aggressive variety of osteosarcoma. The very poor prognosis may be improved with aggressive chemotherapy [3]. This study describes the radiological findings and especially a particular appearance in the initial stage.

Material and methods

Three hundred and seven osteosarcomas, diagnosed and treated between 1969 and 1983 at the Institut Gustave Roussy, were reviewed by two pathologists. The following histological criteria were employed: with low power examination, a cystic appearance is observed, with blood spaces sometimes mimicking an aneurysmal bone cyst. On high power examination, however, the septae contain anaplastic sarcomatous cells, with abundant irregular nuclei and mitoses. The sarcomatous cells appear to be forming mainly osteoid, although it is sometimes difficult to demonstrate such activity. Excluded from the group of telangiectatic osteosarcomas are tumors consisting of more classical osteosarcoma, but containing telangiectatic areas.

Twenty-one tumors were mainly telangiectatic. Seven were excluded: two had had no X-rays before treatment and five were mainly sclerotic on the X-rays and only had a biopsy from the nonsclerotic part of the lesion.

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The 14 remaining cases were studied histologically from material obtained from five biopsies and nine resections. The radiographs were reviewed by two radiologists (DV, ST).

Results

Clinical findings

There were nine males and five females, most aged from 6 to 22 years, but one patient was 71 years old (Table 1). The initial symptom was pain in ten cases, with swelling in seven cases. There were four pathologic fractures, three showed emaciation and in one case (the 71-year-old patient) the tumor was associated with Paget disease. The time between the first symptom and the initial radiological study was less than 2 months in half of the cases, and less than 6 months in all cases.

Radiological findings

Nine cases involved the femur, four the humerus, one the sternum (affected by Paget disease). In the long bones, the lesion was only once located in the shaft and once in the metaphysis (Fig. 5). It was metadiaphyseal in six cases (Fig. 1) and epimetadiaphyseal in five cases (three in the humerus). One lesion of the femur involved the knee joint and the adjacent tibia, but only after a pathological fracture.

All the lesions were purely lytic. The diameter was less than 10 cm in two cases, between 10 and 20 cm in eight cases, and more than 20 cm in four cases. The lesions were central (nine cases) or eccentric (five cases). The appearance of the margins indicates an aggressive tumor, being permeative in eight cases, and better defined (IB or IC) in six cases. In no case was there sclerosis around the lesion (IA).

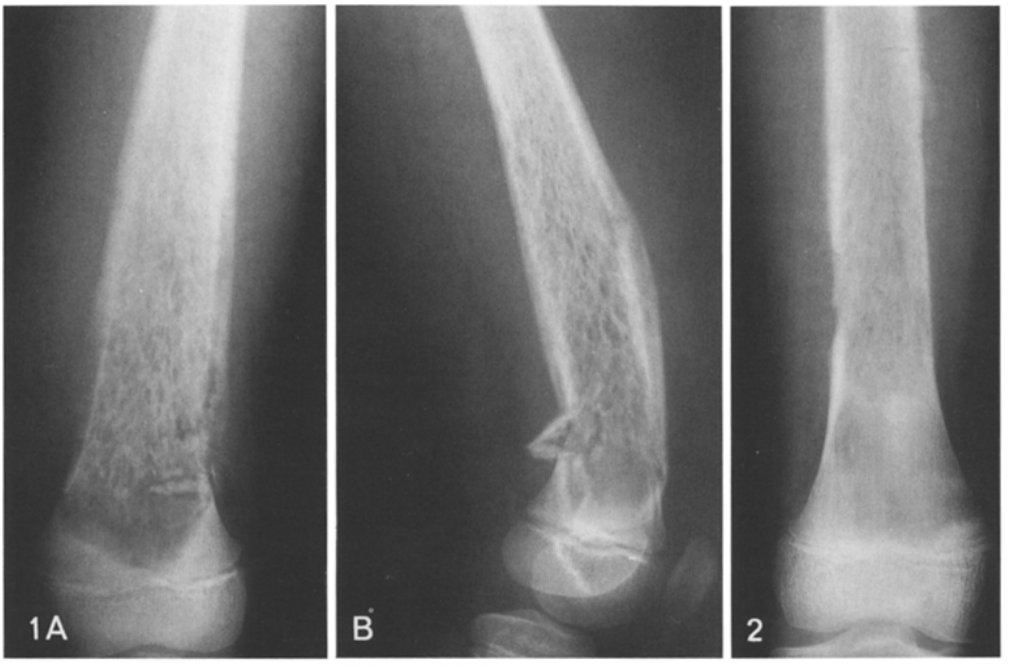
Table 1. Summary of the cases

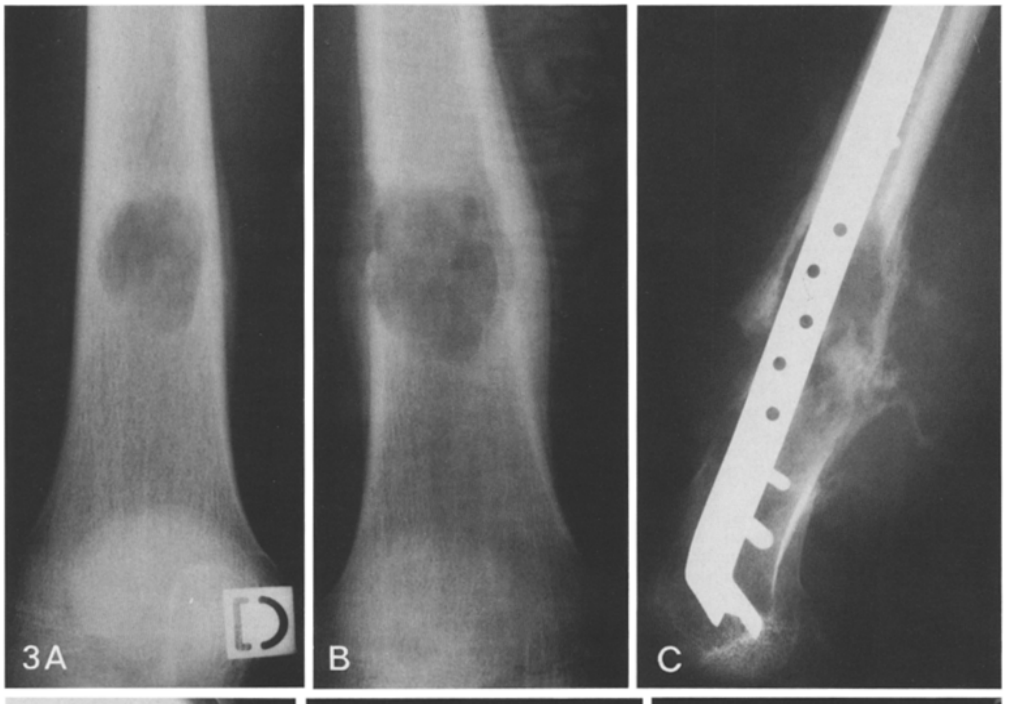
Sex	Age (years)	Site	Initial radiological or histological diagnosis	Size (cm)	Radiological pattern	Figure number	Treatment	Prognosis	Follow-up time
M	13	Distal femur	Simple bone cyst eosinophilic granuloma	14 × 6.5	IB, C, P, ST, F	4	Amputation radiotherapy chemotherapy	Dead	$1\frac{1}{2}$ Years
M	15	Proximal femur	Aneurysmal bone cyst	7 × 4	IB, C		Curettage then amputation radiotherapy chemotherapy	Dead	1 Year
F	15	Distal femur	Osteosynthesis for fracture (correct diagnosis missed)	30 × 15	III, C, P, ST, A, F		Amputation radiotherapy chemotherapy	Lost with widespread metastases	6 Months
M	18	Distal femur		8 × 9	IC, C, P, ST	5	Amputation chemotherapy	Alive	5 Years
F	14	Proximal humerus		23 × 15	parallel striations II, C, P, ST, A	6	Amputation chemotherapy	Lost with widespread metastases	7 Months
F	6	Distal femur		16 × 15	parallel striations III, C, P, JT, F	1	Amputation radiotherapy chemotherapy	Dead	3 Years
M	22	Proximal humerus	Aneurysmal bone cyst giant cell tumor fibrosarcoma Ewing	13 × 10	II, C, P, ST, A, F	7	Conservative surgery chemotherapy	Alive	1 Year
M	18	Proximal humerus		11 × 7	IB, C, P, ST		Radiotherapy amputation	Alive	5 Years
F	11	Femur		32 × 4	III, C, P, ST parallel striations	9	Amputation chemotherapy	Dead	2 Years
M	8	Distal femur		13 × 4	III, C, P, ST parallel striations		Amputation chemotherapy surgery for pulmonary metastasis	Alive	6 Years
M	10	Distal femur		13 × 12	IC, C, P, ST, F		Amputation chemotherapy	Lost with metastases	1 Year
M	14	Proximal humerus	Aneurysmal bone cyst	25 × 15	II, C, F, ST, F		Amputation	Dead	1 Year
F	10	Distal femur		14 × 4	III, C, P, ST		Conservative surgery chemotherapy	Alive	$2\frac{1}{2}$ Years
M	71	Sternum		12 × 6	IC, C, ST, F Paget disease		Radiotherapy	Dead	7 Months

Radiological abbreviations: IB, IC, III: limits, C: Destruction of the cortex, P: periosteal bone formation, ST: Involvement of the soft tissues, A: articular involvement, F: fracture. All lesions are lytic

There was involvement of the soft tissues in 12 cases, destruction of the cortex in 12 cases, and periosteal bone formations in 12 cases (lamellar, perpendicular, or inhomogeneous) with associated Codman's triangles demonstrated in 11 of these. A particular observation was the presence of transient parallel striations (Figs. 1, 2, 6) noted in four cases.

The initial radiological misdiagnoses were aneurysmal bone cyst (Fig. 3) (4), eosinophylic granuloma (1), fibrosarcoma (Fig. 5) (1), and Ewing sarcoma (1).





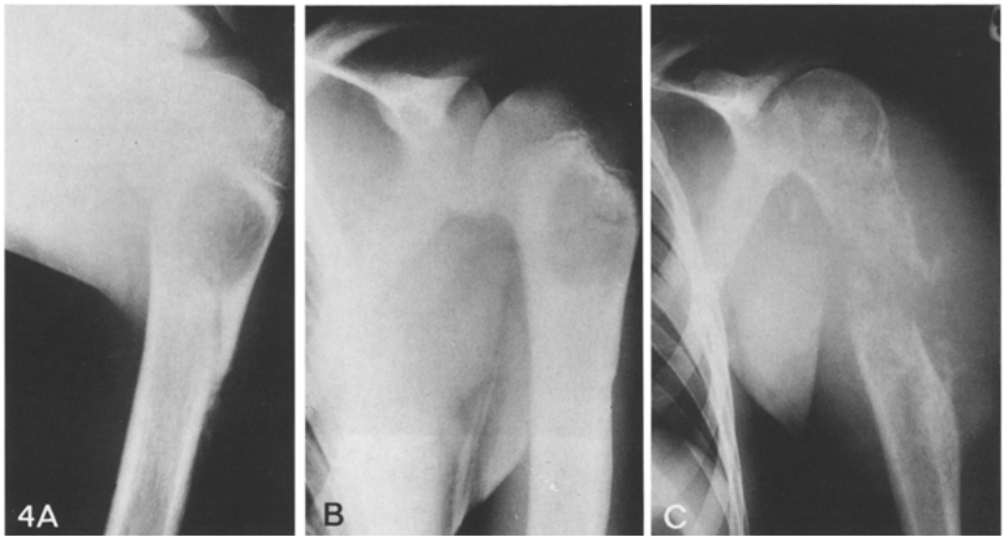
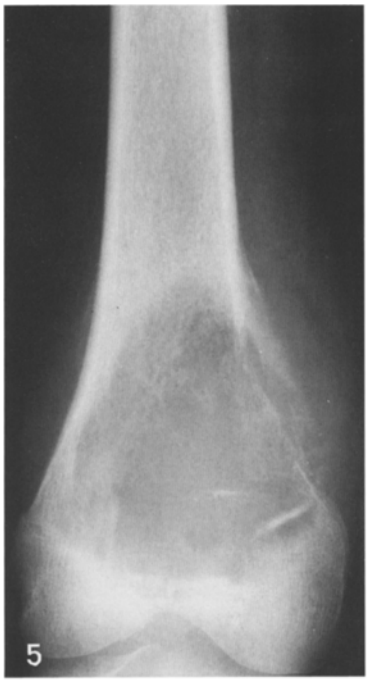


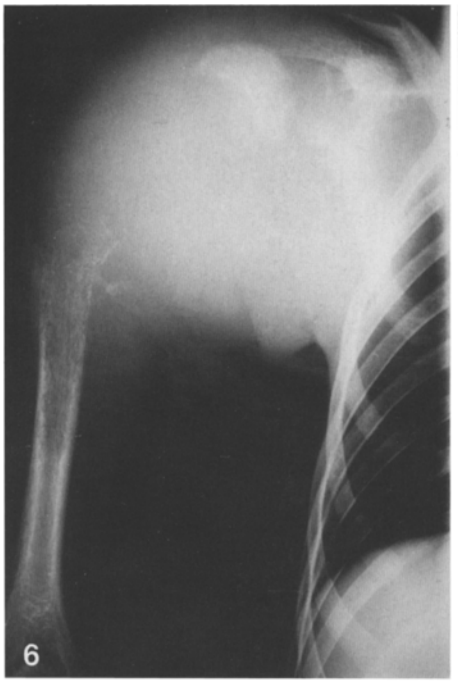
Fig. 1A, B. Six-year-old girl. Characteristic pattern of regular striations in the shaft (AP (**A**) and lateral (**B**) view). The bone is fractured. The tumor involves more than half of bone

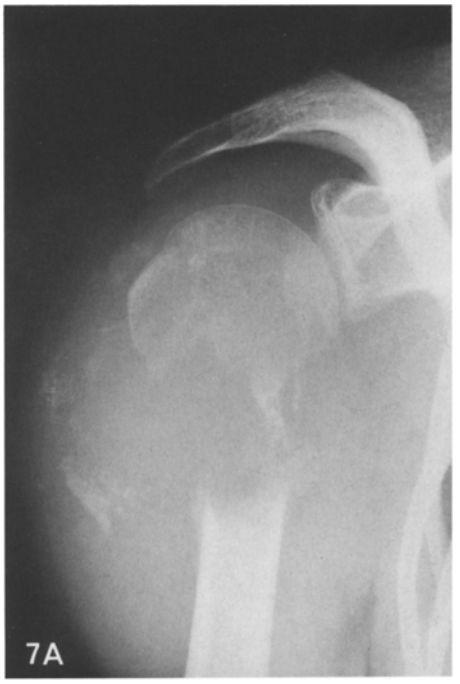
Fig. 2. Eleven-year-old girl. Another typical pattern of regular parallel striations of the shaft

Fig. 3A–C. Nineteen-year-old boy. Routine X-ray examination after an injury reveals a well-defined lytic lesion with lamellar periosteal bone formation, considered to be a bone cyst or an eosinophilic granuloma (**A**). Six months later (**B**) the lesion has increased in size. The histological diagnosis after biopsy was aneurysmal bone cyst. One year later (**C**) pathological fracture had occurred through an aggressive widespread lesion

Fig. 4A–C. Fourteen-year-old girl. Fracture of a probable simple bone cyst (**A**). One month later (**B**) spontaneous regression. No biopsy was performed. Six years later (**C**) an obvious lytic sarcoma







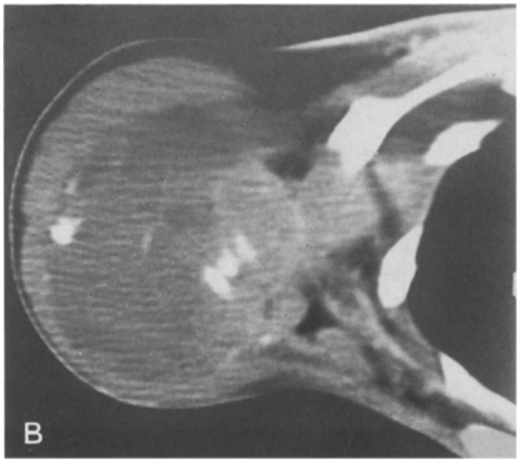


Fig. 5. Eighteen-year-old boy. Lytic tumor with periosteal bone formation and involvement of the soft tissues

Fig. 6. Fourteen-year-old girl. Huge tumor of the proximal humerus. Parallel cortical striations affect the distal part of the humeral shaft

Fig. 7A, B. Lytic tumor of the proximal metaphysis of the humerus (**A**). The involvement of the soft tissues is shown better by CT (**B**)

In summary telangiectatic osteosarcomas radiologically usually are lytic, very aggressive lesions of the long bones (poorly defined, destroying the cortex, with formation of periosteal bone, and invading the soft tissues). The initial pattern of parallel striations is highly suggestive of the diagnosis.

Follow-up

Nine patients died between 7 months and 3 years after diagnosis, five are still alive, three after more than 5 years follow-up, one after two and a half years, and one after one year. Among the four cases with parallel striations, three are dead (indicating no significant prognostic difference).

Discussion

Telangiectatic osteosarcoma is rare, comprising 4.5% of our osteosarcomas (11% for Huvos et al. [3], 2.5% for Matsuno et al. [4]). The difference in incidence can probably be explained by different numbers of simple biopsies or studies of the whole specimen. Pain and swelling are the usual initial symptoms. Males are more often involved (sex ratio 2/1), incidence is greatest in the second decade, and pathological fractures are more frequent than in the common types of osteosarcomas, probably

because the lesion is purely lytic. The time elapsing between the first symptoms and the initial radiological examination is short, often less than 2 months, but that between the radiological study and the establishment of the correct diagnosis may be longer (one year), probably because radiological and histological diagnoses may be difficult. A frequent misdiagnosis is an aneurysmal bone cyst, which is usually better limited and more expanded; the appearance may be similar in the initial stage.

A very special sign is the presence of oblique parallel striations in the shaft (Figs. 1, 2, 6). This finding has already been described twice in the literature [2, 4, 5] but not explained. We propose that this regular oblique striation is due to hypertrophy of the normal structure of the bone. The normal veins of the shaft cross the cortical bone in an oblique direction [1], so we believe that the radiological appearance may be explained by hypertrophy of the normal veins of the bone. This could not be proved as the lesions had already destroyed the cortical bone at the time of the biopsy.

The locations are usual for all osteosarcomas, knee (62%) [3], femur (more than half), and humerus (25%). No periosteal forms have been reported. The usual large size of the lesion and radiological appearance indicate an aggressive lesion (Fig. 6). We had no cases with well-defined margins or sclerosis. Some have been reported [3] resembling benign lesions.

One of our cases was associated with Paget disease; four such others have been reported [3]. One was superimposed on a probably benign lesion which fractured with good spontaneous healing and no biopsy (probably a simple bone cyst), (Fig. 7) and osteosarcoma appearing years later in the same location. We have had few recent cases where chemotherapy was effective. Among the recent cases at the Memorial Hospital [3] the results of chemotherapy have been shown to be at least as good as in the common types of osteosarcoma.

Conclusion

Fourteen cases of telangiectatic osteosarcomas are reported. These are very aggressive lesions, involving mainly the femurs of young adult patients. The presence of a particular radiological appearance at an early stage showing regular parallel striations of the shaft is described.

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