Skeletal Radiol (1985) 14:38–46

Skeletal Radiology

Juxtacortical osteosarcoma: A radiologic and histologic spectrum

E. Levine, M.D., Ph.D.[1], A.A. De Smet, M.D.[1], and M. Huntrakoon, M.D.[2]

Departments of [1]Diagnostic Radiology and [2]Pathology, The University of Kansas College of Health Sciences and Hospital, Kansas City, Kansas, USA

Abstract. Radiologic and pathologic findings were analyzed in four patients with parosteal osteosarcomas, three with periosteal osteosarcomas and one with a high-grade surface osteosarcoma. Plain film and histologic findings considered together are usually distinctive and permit differentiation of these tumors from each other and from other lesions with which they are frequently confused. Prognosis and management are determined by tumor extent and histologic grade. Computed tomography is more accurate than conventional tomography, angiography, and bone scintigraphy for preoperative determination of tumor extent and for assessing tumor relationships to the bone cortex and medullary cavity. Histologically, parosteal osteosarcomas are usually low-grade, while periosteal and high-grade surface osteosarcomas are generally high-grade tumors and have worse prognoses. High-grade surface osteosarcoma, which is indistinguishable in behavior and histology from classical medullary osteosarcoma, requires more aggressive surgical management than parosteal and periosteal osteosarcomas.

Key words: Juxtacortical osteosarcoma – Radiographic features – Pathologic features

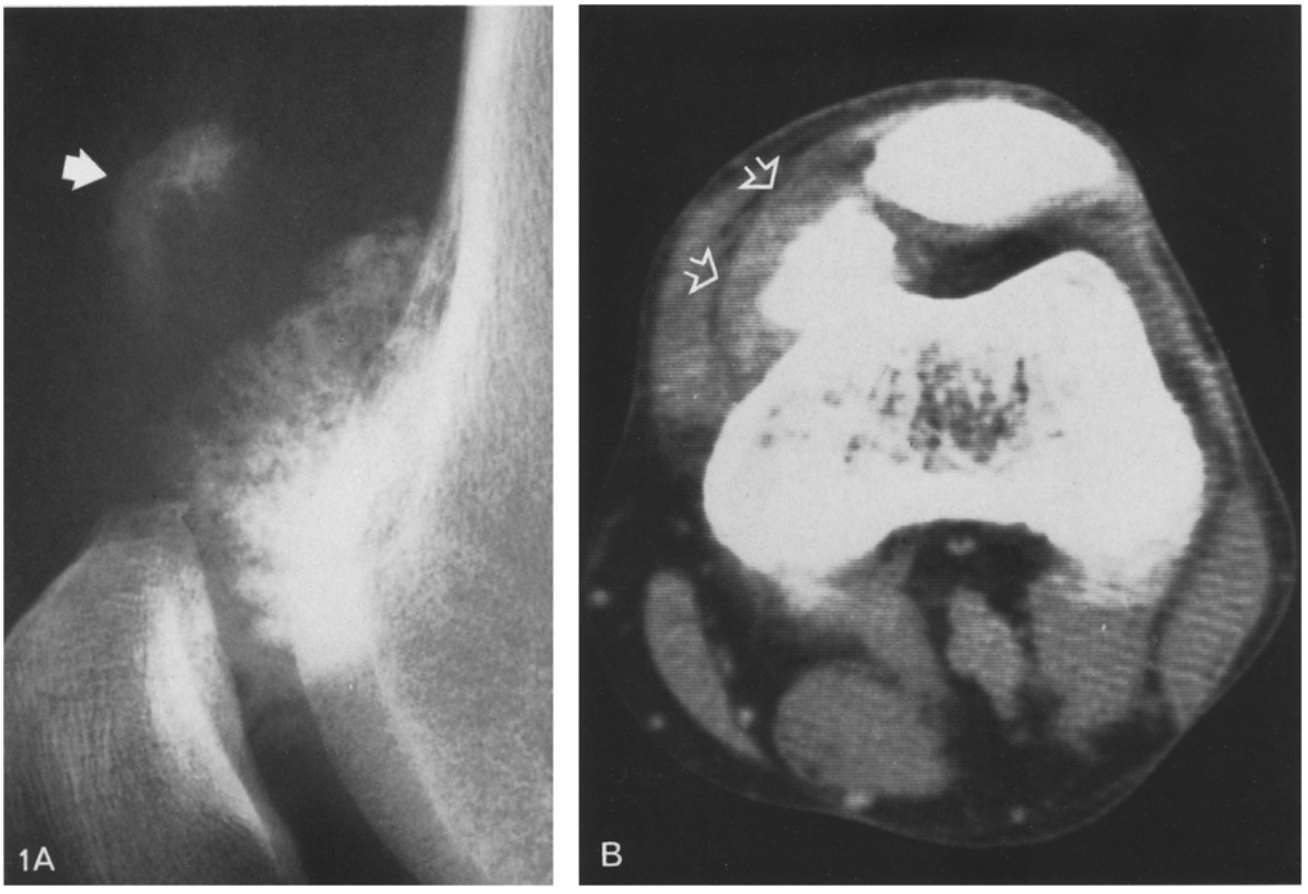
Most osteosarcomas arise within the medullae of long bones and have typical radiologic appearances and rapidly progressive clinical courses [7]. However, osteosarcomas also arise rarely on bone surfaces, and many of these tumors are biologically different from their intramedullary counterparts [7]. In this report, these surface tumors are called "juxtacortical osteosarcomas." Juxtacortical osteosarcoma is generally subdivided into parosteal osteosarcoma and periosteal osteosarcoma [20]. Recently, a third subtype, high-grade surface osteosarcoma, has been recognized as a distinct entity [23]. Because of their rarity, juxtacortical osteosarcomas are frequently misdiagnosed as other lesions such as osteochondromas, myositis ossificans, and chondrosarcomas [3, 10, 22], leading to resection with inadequate surgical margins. In this paper, we analyze the spectrum of radiologic and histologic appearances encountered in juxtacortical osteosarcomas and discuss the use of current imaging techniques including computed tomography (CT), bone scintigraphy, and angiography in their evaluation.

Materials and methods

Of 65 patients with osteosarcomas treated between January 1976 and April 1984, eight (12.3%) had juxtacortical osteosarcomas. There were six men and two women ranging in age from 13 to 39 years (mean age = 29.5 years). Surgical management included wide local (en bloc) tumor excision in seven patients and above-knee amputation in one. Follow-up data were available in all.

All tumors were evaluated by conventional radiographs, hypocycloidal tomography, and bone scintigraphy. Five patients had CT and arteriography of the tumor site. Tumor appearances and extent, determined by these imaging techniques, were analyzed. The intensity of radiophosphate uptake in each tumor was graded relative to sacroiliac joint uptake in posterior views on a scale of 0 to 4+, where 3+ is activity equal to that of the sacroiliac joints [18]. Radiologic and pathologic findings were correlated. The tumors were subdivided into parosteal, periosteal, and high-grade surface osteosarcomas using established radiologic and histologic criteria [20, 21, 23]. Tumor histologic differentiation was graded, using previously described criteria [1, 3, 23], on a scale of 1 to 3, where grade 1 indicates a well-differentiated tumor and grade 3 designates an anaplastic poorly differentiated lesion.

Address reprint requests to: Errol Levine, M.D., Ph.D., Department of Diagnostic Radiology, 2169 Bell Memorial Hospital, The University of Kansas College of Health Sciences and Hospital, Rainbow Boulevard at 39th Street, Kansas City, KS 66103, USA



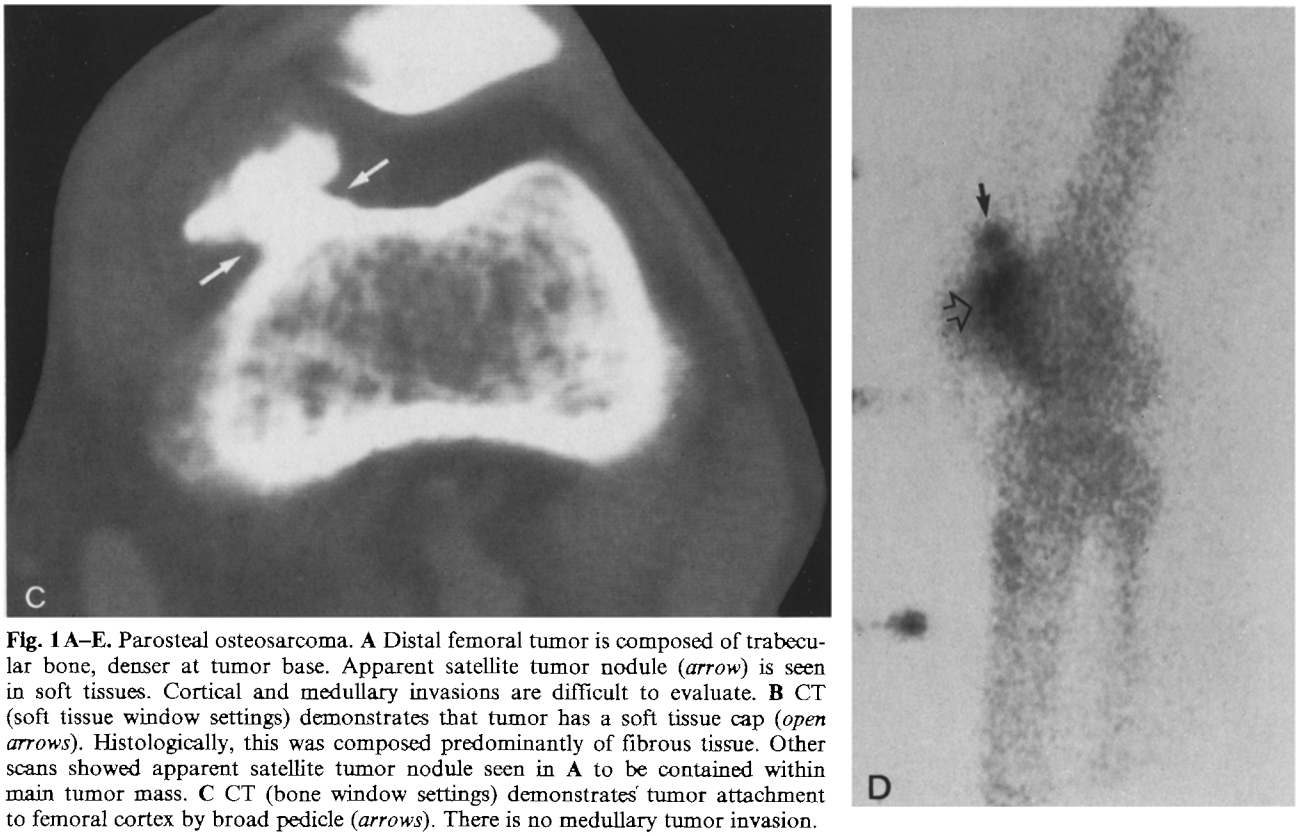


Fig. 1 A–E. Parosteal osteosarcoma. **A** Distal femoral tumor is composed of trabecular bone, denser at tumor base. Apparent satellite tumor nodule (*arrow*) is seen in soft tissues. Cortical and medullary invasions are difficult to evaluate. **B** CT (soft tissue window settings) demonstrates that tumor has a soft tissue cap (*open arrows*). Histologically, this was composed predominantly of fibrous tissue. Other scans showed apparent satellite tumor nodule seen in **A** to be contained within main tumor mass. **C** CT (bone window settings) demonstrates tumor attachment to femoral cortex by broad pedicle (*arrows*). There is no medullary tumor invasion. **D** Lateral radiophosphate scintigram displays increased tracer uptake in tumor (*open arrow*) and in small "satellite" tumor nodule (*arrow*). Ten cm markers are present anteriorly (**E** on page 40)

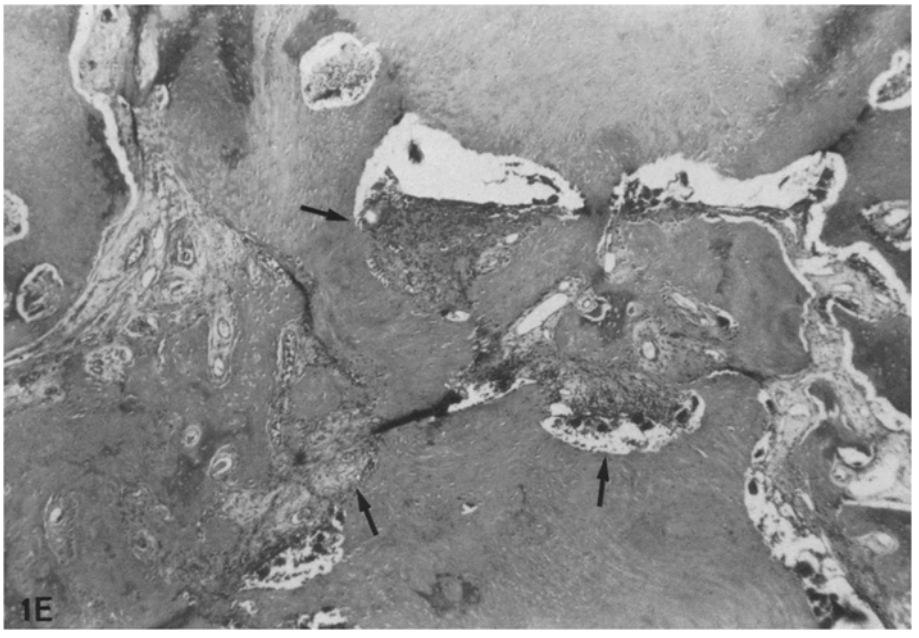
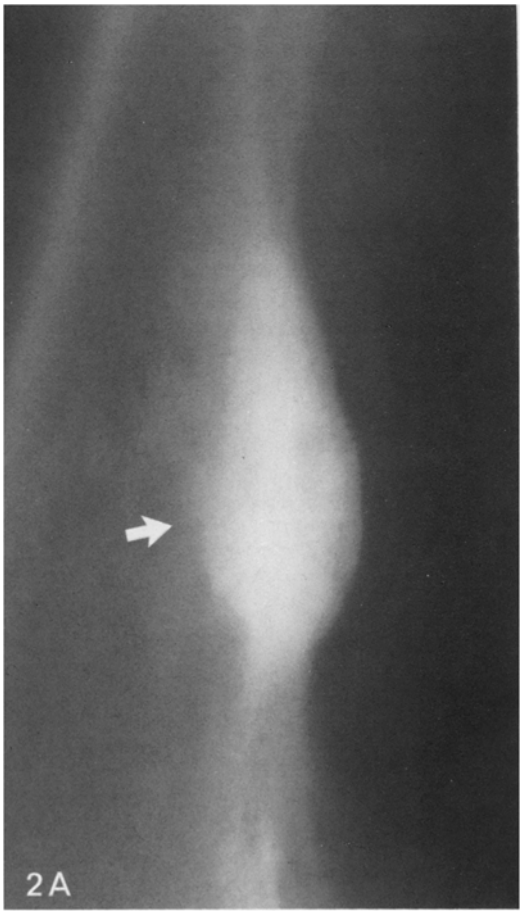


Fig. 1E. Miroscopically, (H and E ×16), tumor is composed of irregular bone trabeculae with interspersed foci of spindle cells (*arrows*). At high power, spindle cells showed mild atypia and occasional mitotic figures



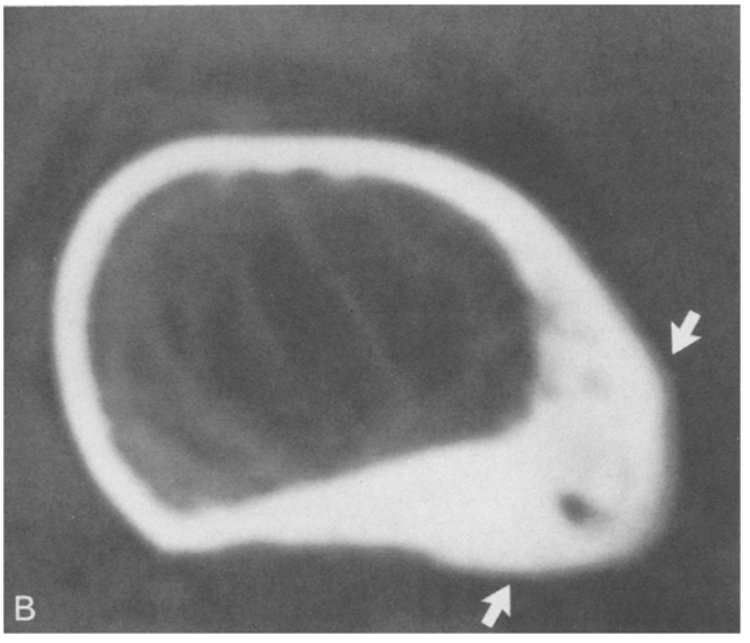


Fig. 2A, B. Parosteal osteosarcoma. **A** On lateral tomography of distal femur, dense tumor apparently extends into medullary cavity anteriorly (*arrow*). **B** On CT, tumor (*arrows*) cannot be distinguished from adjacent thickened bone cortex, but there is no medullary tumor extension. Note small lucencies within dense tumor

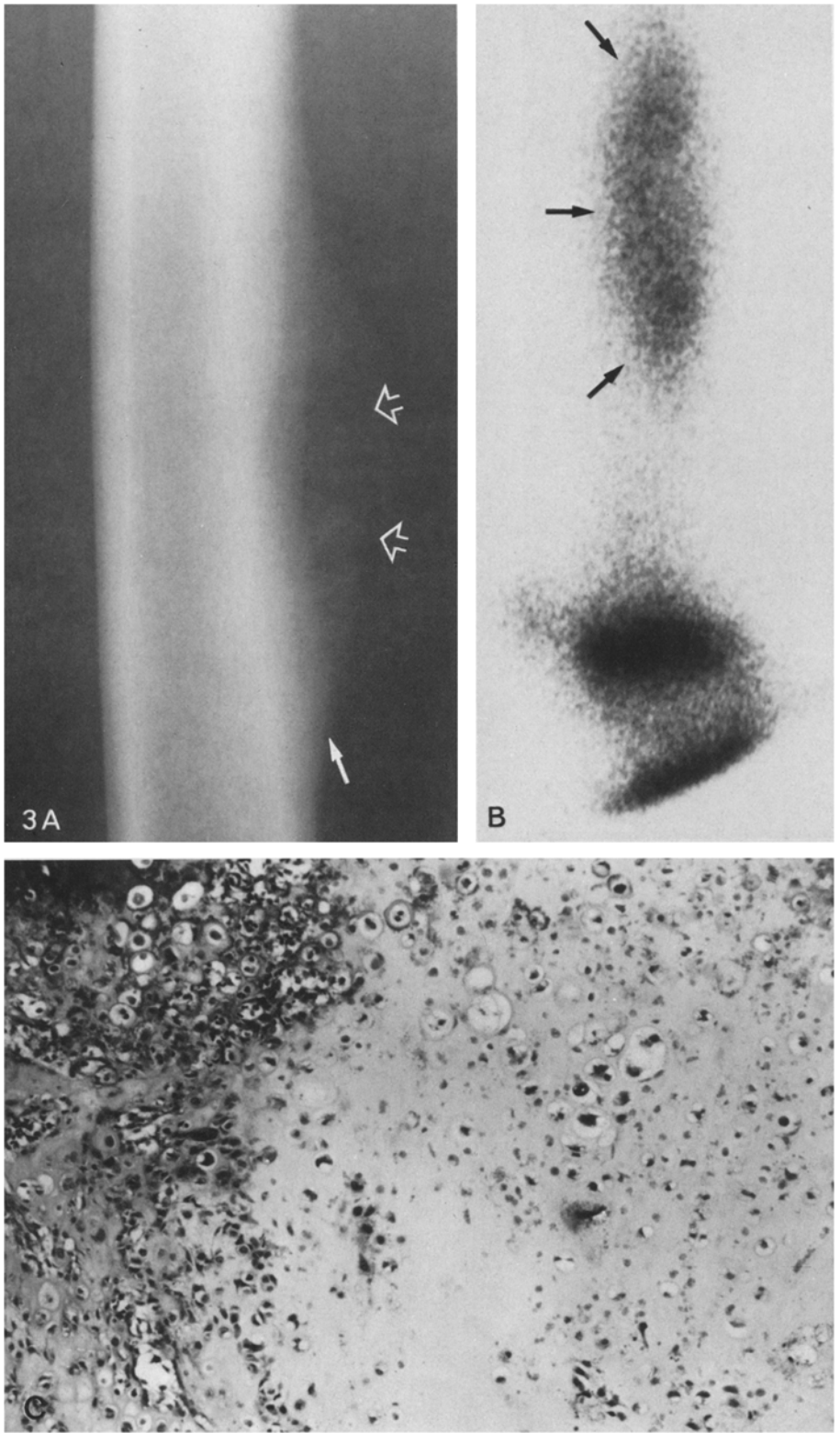


Fig. 3A–C. Periosteal osteosarcoma
A Lateral projection of distal femoral diaphysis demonstrates small soft tissue mass (*open arrows*) containing delicate bone spicules. Tumor produces shallow cortical erosion posteriorly, but endosteal cortex is preserved. *Arrow* indicates periosteal new bone formation
B Lateral radiophosphate scintigram displays increased tracer activity extending to anterior femoral surface (*arrows*), incorrectly suggesting medullary tumor extension
C Microscopically, (H and E ×40), tumor is composed predominantly of lobules of moderately differentiated malignant cartilage cells with peripheral hypercellularity. At high power, these hypercellular zones contained spindle cells and fine, lace-like osteoid

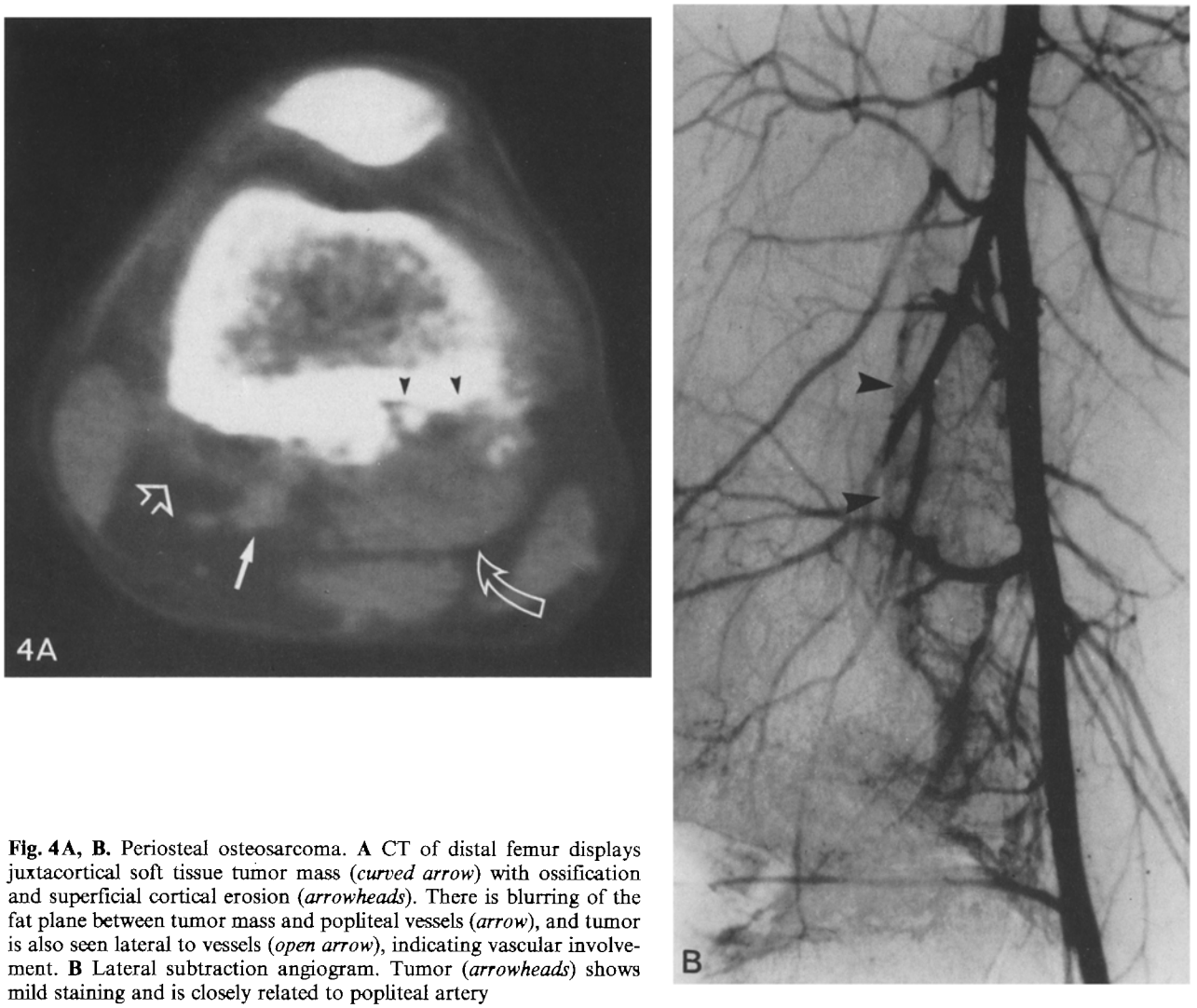


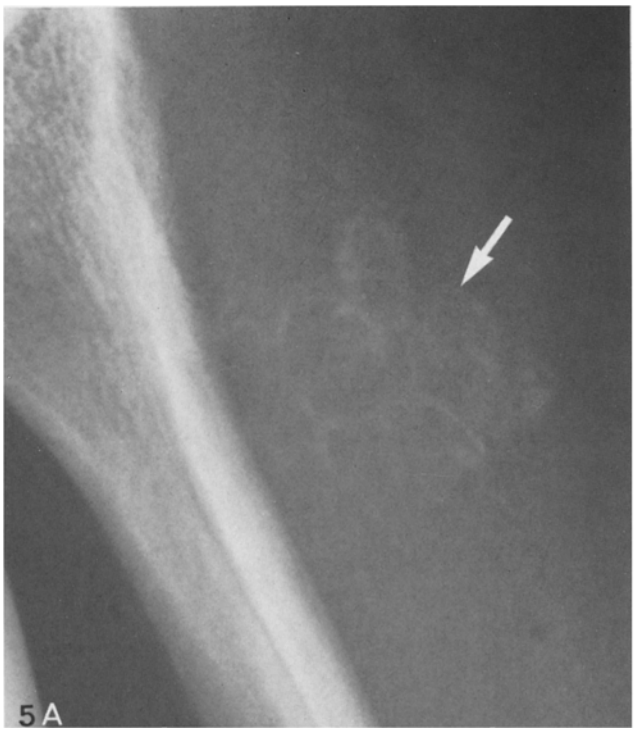
Fig. 4A, B. Periosteal osteosarcoma. **A** CT of distal femur displays juxtacortical soft tissue tumor mass (*curved arrow*) with ossification and superficial cortical erosion (*arrowheads*). There is blurring of the fat plane between tumor mass and popliteal vessels (*arrow*), and tumor is also seen lateral to vessels (*open arrow*), indicating vascular involvement. **B** Lateral subtraction angiogram. Tumor (*arrowheads*) shows mild staining and is closely related to popliteal artery

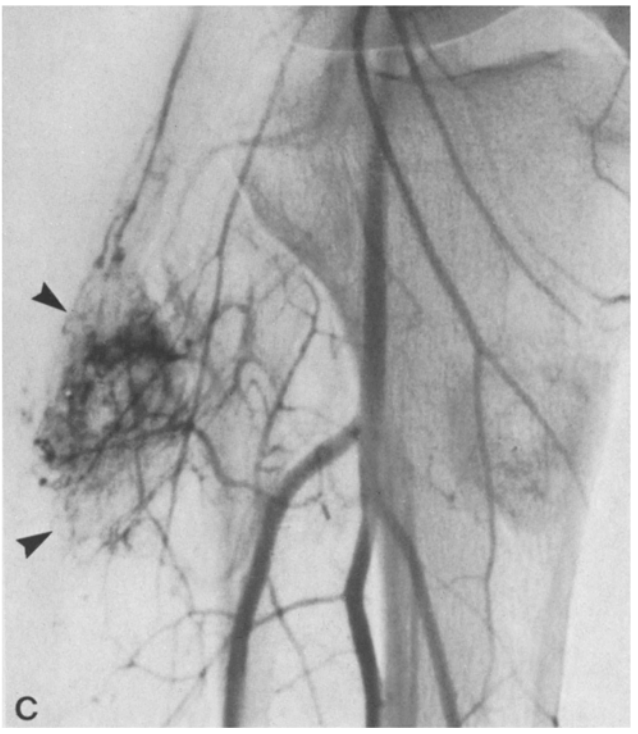
Results

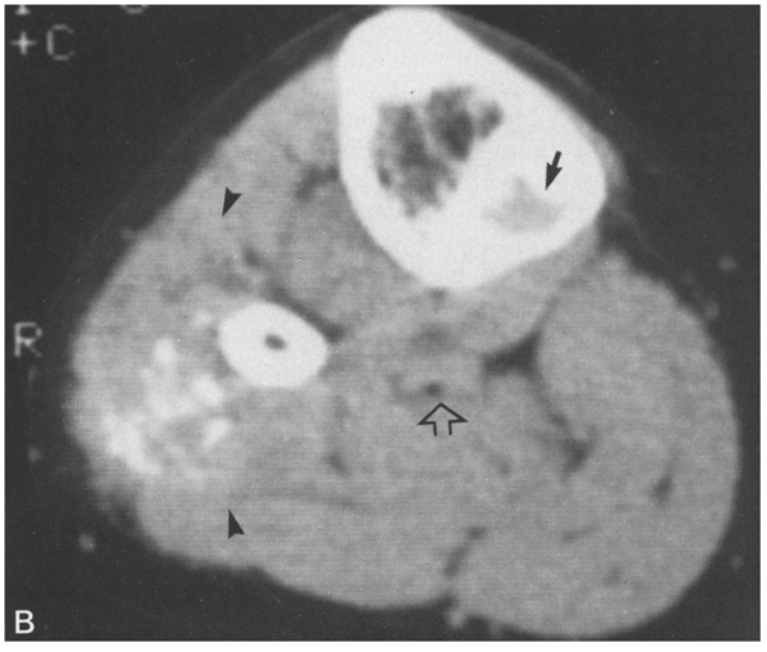
Parosteal osteosarcomas

Four patients, all men, had parosteal osteosarcomas. They were aged 24, 29, 37, and 39 years, respectively. Two tumors arose from the distal femur, one from the proximal tibia, and one from the tibial midshaft. One tumor (Fig. 1) represented a recurrence 11 years after local resection of an "osteochondroma." The tumors appeared on plain films as juxtacortical masses of varying densities. Three were composed of trabecular bone (Fig. 1), and one was composed of dense, compact, ivory-like bone (Fig. 2). All four tumors showed lucent areas within the otherwise dense tumor masses (Figs. 1 and 2). Three were denser at their bases than at their peripheries (Fig. 1), and two exhibited thin, radiolucent clefts between parts of the tumors and the underlying cortex. However, at least part of the tumor was inseparable from the underlying cortex in all patients so that cortical and medullary tumor invasions were difficult to evaluate on plain films and conventional tomography (Figs. 1 and 2). Thickening of the adjacent cortex was observed in two patients.

CT scans were available in three patients and showed no vascular or medullary tumor invasion (Figs. 1 and 2). However, tumor bone could not be distinguished from thickened, adjacent cortex (Fig. 2). Radiophosphate uptake corresponded to anatomic tumor dimensions in all patients (Fig. 1), and intensity of uptake was graded 4+ in two scans, 3+ in one, and 1+ in one. Angiography was performed in one patient and showed an avascular tumor. No patient showed medullary tumor invasion on pathologic examination. Histologically, the tumors were considered grade 1 lesions [1]. They were characterized by abundant, well-







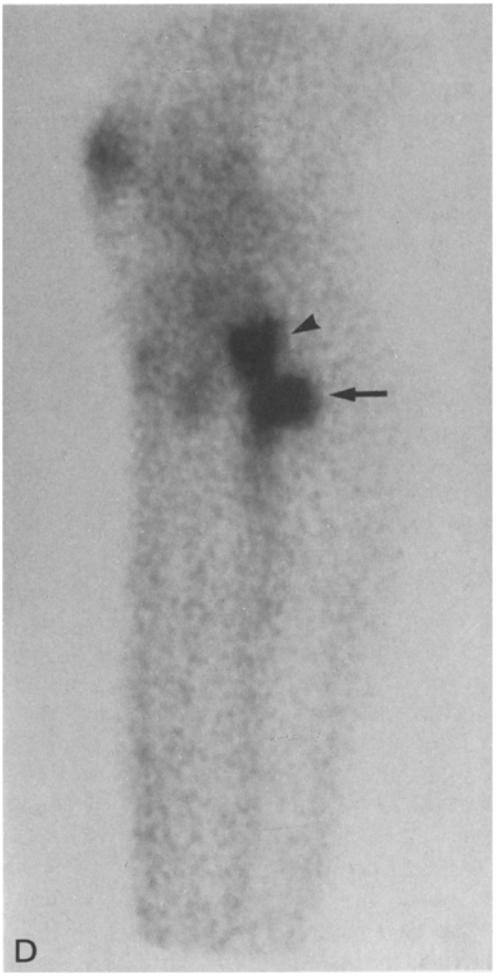


Fig. 5A–E. High-grade surface osteosarcoma. **A** Lateral view shows partially ossified soft tissue mass (*arrow*) related to uneroded posterior fibular cortex. **B** CT displays partially ossified, ill-defined soft tissue mass (*arrowheads*), which is infiltrating adjacent muscles but is remote from popliteal vessels (*open arrow*). Monostotic fibrous dysplasia involves tibial cortex (*arrow*). **C** Anteroposterior subtraction angiographic study demonstrates tumor neovascularity and hypervascularity (*arrowheads*). Early venous drainage was evident in later films. **D** Lateral radiophosphate scintigram displays intense (3+) tracer uptake in soft tissue mass (*arrow*). There is also increased tracer uptake in proximal fibula (*arrowhead*), incorrectly suggesting bone invasion by tumor (**E** on page 44).

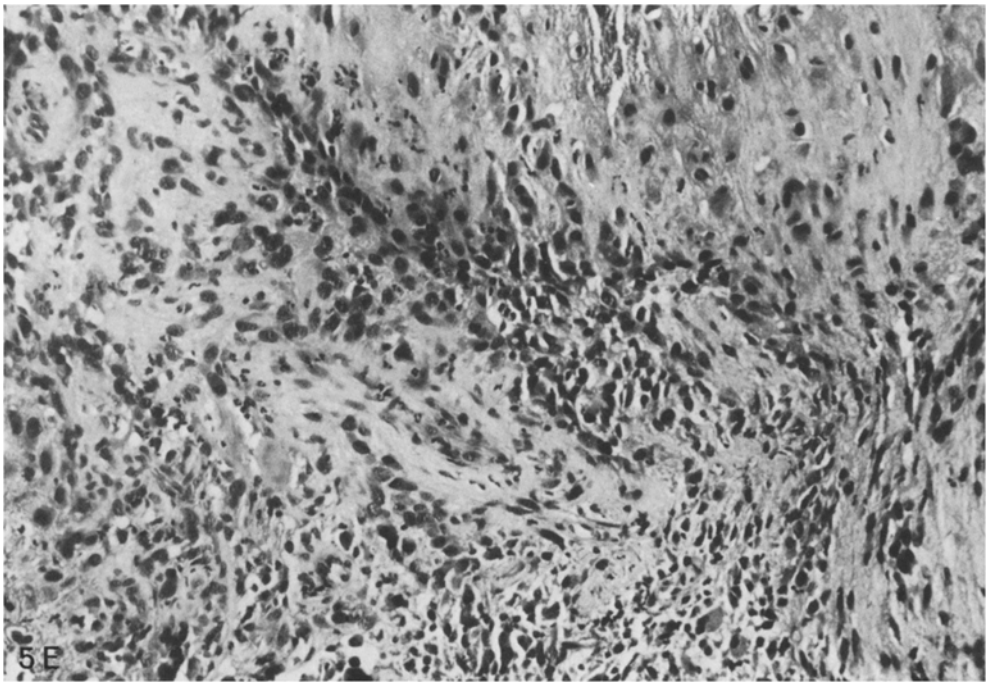


Fig. 5E. Microscopically, (H and E ×40), tumor is composed of highly anaplastic cells among which osteoid is evident

formed osseous trabeculae between which lay foci of spindle cells exhibiting mild atypia and occasional mitotic figures (Fig. 1). No patient had metastases at presentation, and neither local tumor recurrences nor metastases were observed during postsurgical follow-up periods of 7, 11, 36, and 63 months, respectively.

Periosteal osteosarcomas

Three patients, two males and one female, aged, respectively 13, 22, and 39 years, had distal femoral periosteal osteosarcomas. One tumor represented a recurrence nine years after local resection. On plain films and conventional tomography, the tumors presented as juxtacortical soft tissue masses containing small bone spicules (Fig. 3). Periosteal new bone formation and superficial cortical erosion (Fig. 3) were seen in two patients, while neither finding was noted in the third. Angiography showed that one tumor was avascular and that two were hypovascular (Fig. 4). CT was available in one patient and demonstrated vascular invasion (Fig. 4). Since the tumor could not be dissected from the vessels at surgery, an above-knee amputation was performed. The other two patients had wide local tumor excisions.

Radiophosphate uptake intensity was graded 3+ in two tumors and 2+ in one. The extent of osseous tracer activity incorrectly suggested medullary tumor invasion in all three patients (Fig. 3). One patient also showed an "extended uptake pattern" [19] with increased activity involving the knee joint and proximal tibia, in the absence of tumor involvement at these sites. No patient had medullary tumor invasion on pathologic examination, although superficial cortical erosion was evident in two, as predicted radiologically. Histologically, the tumors were considered grade 2 lesions [3, 20] and consisted of lobules of moderately differentiated cartilage cells separated by septa containing spindle cells and osteoid (Fig. 3). No patient had metastases at presentation and neither local tumor recurrences nor metastases were observed during postsurgical follow-up periods of 81, 97, and 98 months, respectively.

High-grade surface osteosarcoma

One patient, a 33-year-old woman, had a high-grade surface osteosarcoma arising from the posterolateral aspect of the fibular neck. Its radiologic features are illustrated (Fig. 5). Bone scintigraphy erroneously suggested fibular invasion (Fig. 5). Pathologic examination confirmed the radiologic findings of a soft tissue mass without erosion of the underlying cortex. Histologically, the tumor was grade 3 and was composed of anaplastic cells among which osteoid could be identified (Fig. 5). No metastases were detected at presentation, and

the patient has no evidence of local tumor recurrence or of metastases during a 14-month postsurgical follow-up period.

Discussion

Parosteal osteosarcoma is a rare, slow-growing, bone-producing neoplasm, comprising slightly less than 4% of osteosarcomas [5]. The tumor has a peak incidence in the third and fourth decades [11] and most commonly arises from the popliteal surface of the distal femur [21]. Most tumors present characteristic plain film appearances. They are usually firmly attached to the adjacent, often thickened, bone cortex by a broad base [21] and frequently cannot be distinguished from it by either radiography or CT (Fig. 2). As the tumor grows, it encircles the shaft, often leaving a thin, lucent cleft between the underlying bone and the neoplasm [21]. Periosteal new bone formation is rarely found [5]. Even dense tumors contain small lucencies (Fig. 2) [24] which may represent fibrous or cartilaginous tissue, normal fat, entrapped benign soft tissues or dedifferentiated (high grade) areas of osteosarcoma [4, 24].

Preoperative assessment of tumor extent is important for prognosis and treatment planning [4]. CT is the most effective method for delineating the relationship of the ossified mass to adjacent muscles and vessels and for determining whether medullary invasion has occurred [4, 9], although small areas of medullary invasion may be difficult to detect on CT [16]. Conventional tomography is not useful for these purposes and, since most parosteal osteosarcomas are avascular or hypovascular [17, 25], angiography is also of limited use for determining tumor extent. Bone scintigraphy does not add to the information provided by plain films and CT.

Most parosteal osteosarcomas are histologically low-grade lesions [21], although some tumors contain high-grade (dedifferentiated) areas [1, 24]. The histologic appearances are distinctive (Fig. 1). Some authors have described two distinctly different forms of parosteal osteosarcoma [15]. They claim that the predominant type is originally a low-grade malignant tumor which metastasizes after a long symptom-free interval, while the other type is a highly malignant tumor from the outset. However, it seems more appropriate that the latter be categorized as high-grade surface osteosarcomas [23]. The prognosis of parosteal osteosarcoma is better than that of medullary osteosarcoma [21] and depends on tumor grade [1, 4] and whether medullary invasion has occurred [4, 24]. Metastases occur mainly with high-grade tumors that have invaded the medullary cavity [4]. Well-differentiated low-grade tumors are sometimes called parosteal "osteomas" [6, 14]. However, if incompletely resected, such tumors often recur, sometimes in a dedifferentiated form, and metastases may occur [24]. Accordingly, appropriate therapy requires complete tumor removal by wide local (en bloc) excision [4, 12], regardless of histologic grade [24].

Differentiation of parosteal osteosarcoma from myositis ossificans circumscripta and osteochondroma is important clinically. Myositis ossificans, which may be treated conservatively or by limited marginal excision [2, 26], presents radiologically as an ossified mass adjacent to bone. It is differentiated from parosteal osteosarcoma by its lack of attachment to underlying bone and by the presence of peripheral mineralization and a lucent center on CT [2, 26]. An osteochondroma arises from the underlying bone with its medullary cavity and cortex continuous with those of the affected bone [5]. On the other hand, a cortex is present beneath a parosteal osteosarcoma which, therefore, does not share a common medullary cavity with the underlying bone [5].

Periosteal osteosarcoma is a less common juxtacortical tumor, accounting for about 1% of osteosarcomas [5]. Most lesions occur in the second and third decades [20] with the femur and tibia being the most common locations [8, 20]. The plain film finding of a juxtacortical soft tissue mass containing fine bone spicules and associated with periosteal new bone formation and superficial cortical erosion is highly suggestive of periosteal osteosarcoma [8, 20]. The medullary cavity is usually spared [8, 20]. CT is useful for determining the relationship of the tumor to major vessels and to the bone cortex (Fig. 4) [9]. Conventional tomography does not usually help in further evaluation. Radiophosphate scans often incorrectly suggest medullary invasion (Fig. 3). Since most tumors are avascular or hypovascular, angiography is of limited use in determining tumor extent (Fig. 4).

Periosteal osteosarcomas are generally high-grade (grade 2 or 3), chondroblastic osteosarcomas and differ strikingly in their histologic appearances from parosteal osteosarcomas (Fig. 3) [3, 20]. Although the predominance of malignant cartilage may suggest a diagnosis of chondrosarcoma [3], the presence of osteoid indicates the true nature of the tumor [20]. The general prognosis is better than for medullary osteosarcoma but not as good as for parosteal osteosarcoma [20]. Wide local tumor excision, if technically feasible, and adjuvant

chemotherapy are the generally recommended forms of management [13, 20].

High grade surface osteosarcomas are rare juxtacortical tumors, accounting for less than 1% of osteosarcomas [23]. Most tumors occur in the femur where they present similar radiologic appearances to those of periosteal osteosarcoma [23]. CT helps determine tumor extent and relationships to adjoining structures including blood vessels, muscles, and bone cortex. Radiophosphate scans may suggest osseous involvement in the absence of cortical erosion (Fig. 5). Angiography suggests the high-grade nature of the tumor (Fig. 5) by showing findings similar to those in medullary osteosarcoma [17, 25]. Histologically, high-grade surface osteosarcoma is less differentiated than periosteal osteosarcoma but is indistinguishable from intramedullary osteosarcoma and has the same poor prognosis [23]. Accordingly, appropriate management of high-grade surface osteosarcoma requires radical local excision and adjuvant chemotherapy [12, 13].

In conclusion, juxtacortical osteosarcomas present an interesting spectrum of radiologic and histologic appearances. The plain film and histologic findings considered in combination are distinctive and permit differentiation of the three subtypes of juxtacortical osteosarcoma from each other and from other lesions with which they are frequently confused. As with other musculoskeletal tumors, accurate anatomic staging is an essential part of preoperative treatment planning [12]. Radiologic evaluation of these neoplasms should consist of plain films and CT. Angiography may occasionally be necessary in treatment planning to locate major vessels and exclude vascular encasement if CT does not clarify this issue. Conventional tomography and bone scintigraphy do not aid in diagnosis or determination of disease extent. In fact, they may be misleading (Figs. 2, 3, and 5).

References

1. Ahuja SC, Villacin AB, Smith J, Bullough PG, Huvos AG, Marcove RC (1977) Juxtacortical (parosteal) osteogenic sarcoma. Histological grading and prognosis. J Bone Joint Surg [Am] 59:632
2. Amendola MA, Glazer GM, Agha FP, Francis IR, Weatherbee L, Martel W (1983) Myositis ossificans circumscripta: computed tomographic diagnosis. Radiology 149:775
3. Bertoni F, Boriani S, Laus M, Campanacci M (1982) Periosteal chondrosarcoma and periosteal osteosarcoma. Two distinct entities. J Bone Joint Surg [Br] 64:370
4. Campanacci M, Picci P, Gherlinzoni F, Guerra A, Bertoni F, Neff JR (1984) Parosteal osteosarcoma. J Bone Joint Surg [Br] 66:313
5. Dahlin DC (1978) Bone tumors; general aspects and data on 6,221 cases, 3rd edn. Charles C Thomas, Springfield, Illinois, p 250
6. Dahlin DC (1978) Bone tumors: general aspects and data on 6,221 cases, 3rd edn. Charles C Thomas, Springfield, Illinois, p 71
7. Dahlin DC, Unni KK (1977) Osteosarcoma of bone and its important recognizable varieties. Am J Surg Pathol 1:61
8. deSantos LA, Murray JA, Finklestein JB, Spjut HJ, Ayala AG (1978) The radiographic spectrum of periosteal osteosarcoma. Radiology 127:123
9. Destouet JM, Gilula LA, Murphy WA (1979) Computed tomography of long-bone osteosarcoma. Radiology 131:439
10. Dwinnell LA, Dahlin DC; Ghormley RK (1954) Parosteal (juxtacortical) osteogenic sarcoma. J Bone Joint Surg [Am] 36:732
11. Edeiken J, Farrell C, Ackerman LV, Spjut HJ (1971) Parosteal osteosarcoma. AJR 111:579
12. Enneking WF (1983) Musculoskeletal tumor surgery, 1st edn. Churchill Livingstone, New York, p 69
13. Enneking WF (1983) Musculoskeletal tumor surgery, 1st edn. Churchill Livingstone, New York, p 1108
14. Geschickter CF, Copeland MM (1951) Parosteal osteoma of bone: a new entity. Ann Surg 133:790
15. Lorentzon R, Larsson SE, Boquist L (1980) Parosteal (juxtacortical) osteosarcoma. J Bone Joint Surg [Br] 62:86
16. Orcutt J, Ragsdale BD, Curtis DJ, Levine MI (1981) Misleading CT in parosteal osteosarcoma. AJR 136:1233
17. Probst FP (1971) Angiography in juxtacortical osteosarcomas. Case report with special reference to the differential diagnosis. Acta Radiol [Diagn] (Stockh) 11:49
18. Simon MA, Kirchner PT (1980) Scintigraphic evaluation of primary bone tumors. J Bone Joint Surg [Am] 62:758
19. Thrall JH, Geslien GE, Corcoron RJ, Johnson MC (1975) Abnormal radionuclide deposition patterns adjacent to focal skeletal lesions. Radiology 115:659
20. Unni KK, Dahlin DC, Beabout JW (1976) Periosteal osteogenic sarcoma. Cancer 37:2476
21. Unni KK, Dahlin DC, Beabout JW, Ivins JC (1976) Parosteal osteogenic sarcoma. Cancer 37:2466
22. Van Der Heul RO, Von Ronnen JR (1967) Juxtacortical osteosarcoma. Diagnosis, differential diagnosis, treatment, and an analysis of eighty cases. J Bone Joint Surg [Am] 49:415
23. Wold LE, Unni KK, Beabout JW, Pritchard DJ (1984) High-grade surface osteosarcomas. Am J Surg Pathol 8:181
24. Wold LE, Unni KK, Beabout JW, Sim FH, Dahlin DC (1984) Dedifferentiated parosteal osteosarcoma. J Bone Joint Surg [Am] 66:53
25. Yaghmai I (1971) Angiographic features of osteosarcoma. AJR 129:1073
26. Zeanah WR, Hudson TM (1982) Myositis ossificans. Radiologic evaluation of two cases with diagnostic computed tomograms. Clin Orthop 168:187