Parosteal osteosarcoma

2–23-year follow-up of 33 patients

Peter Ritschl[1], Christian Wurnig[1], Gerhard Lechner[3] and Alfred Roessner[2]

In a study of the working group for bone tumors of the German Orthopedic Society, 33 patients with a histologically confirmed parosteal osteosarcoma at reexamination underwent clinical and radiographic follow-up. Local recurrence occurred in all the cases after intralesional surgery and in 4 of 8 cases after marginal excision. The grade of differentiation was decisive for the prognosis. Despite intralesional surgery, the prognosis for Grade 1 tumors was good. Metastases developed in 1 of 23 patients with a Grade 1 tumor and in 4 of 9 patients with a Grade 2 tumor. The single patient with a Grade 3 tumor was treated with adjuvant chemotherapy and was free of disease after 5 years.

[1]Orthopedic Department, University of Vienna, [2]Gerhard-Dogmagk-Institute for Pathology, Westphalien Wilhelm-University, Münster, Germany, and [3]X-Ray Department of the First Surgical Clinic, University of Vienna, Vienna, Austria
Submitted 90-05-27. Accepted 91-02-02

Parosteal osteosarcoma is regarded as a distinct entity with good prognosis as compared with classic osteosarcoma (Campanacci et al. 1984, Dahlin and Unni 1986, Geschickter and Copeland 1951, Scaglietti and Calandriello 1962, Stevens et al. 1957). In a retrospective multicenter study of parosteal osteosarcomas, local and distant recurrence was evaluated and related to surgical margins and histologic grade.

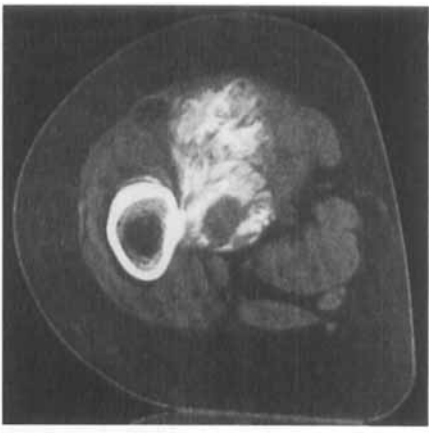
Patients and methods

Twelve departments with a total of 45 patients treated between 1965 through 1985 took part in the study (Table 1). The radiographs, histologic slides (33 patients), and clinical findings were evaluated retrospectively. The median age was 34 (12–72) years and the male to female rate 2:3. Twenty-three of the tumors were located in the distal femur, 19 of them in the popliteal region.

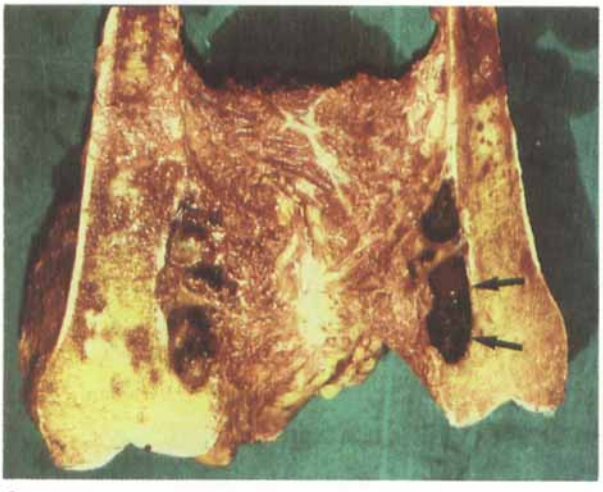
Table 1. The working group for bone tumors of the German Orthopedic Society and number of cases submitted by the participating departments

	Evaluated cases	
	Clinical and radiographic	Histologic
Germany		
Westphalien Wilhelm-University, Münster (Paulus Wuisman)	9	8
Orthopedic Department, University of Heidelberg (Arnim Braun)	5	–
Orthopedic Clinic, Volmarstein (Wolfgang Becker)	3	3
Orthopedic Department and Clinic, University of Berlin (Andreas Kefenbaum)	1	1
Orthopedic Department and Clinic, Düsseldorf (Winfried Winkelmann)	1	–
Orthopedic Department and Clinic, University of München (Rudolf Gradinger)	2	–
Klinikum, University of Ulm (Wolf Mutschler)	2	2
Ingolstadt (Hans Jörg Gubba)	1	–
Austria		
Orthopedic Clinic, University of Vienna (Peter Ritschl)	7	7
Orthopedic Clinic, University of Innsbruck (Wolfgang Russe)	5	5
Surgical Clinic, University of Graz (Richard Trauner)	2	–
CSFR		
Orthopedic Department, University of Prague (Zdenek Matejovsky)	7	7
Total	45	33

A



B



C

Figure 1. Case 16, a 30-year-old female.

A. Parosteal osteosarcoma of the distal femur. Probable gap between the tumor and proximal cortical bone. Penetration into the medullary cavity cannot be verified by a plain radiograph.

B. Computed tomogram, hyperintense and hypointense areas that encircle the bone about two thirds of its circumference. An intramedullary invasion cannot be observed on this slice.

C. Cut specimen after wide resection. In addition to solid tumor areas, there are also large regressive cysts. Arrows show intramedullary extension. Condition after polychemotherapy COSS 86, grade of regression 6 according to Salzer-Kuntschik (1983).

Twenty patients had been operated on before referral to the tumor center, and 25 were referred without previous surgery. The average length of time from the first operation until treatment at the tumor center was 3 (0.2–15) years. Of the 20 patients referred after surgery, 15 had been operated on once, 4 twice, and 1 three times. The interventions were five incisional biopsies, 19 resections, and two amputations. The resections and amputations were all intralesional. The initial histologic diagnoses included osteochondroma, myositis ossificans, ectopic metaplastic bony tissue, desmoplastic fibroma, chronic inflammation or osteitis, and osteoid osteoma. In 10 cases the diagnosis was sarcoma, in 7 of them parosteal osteosarcoma.

Due to diagnostic difficulties, 3 patients had two intralesional resections, each at the tumor center. Treatment after correct diagnosis by biopsy comprised 31 resections (Figure 1), 12 amputations, and two rotation plasties (Figure 2). In all 14 cases of amputation and rotation plasty, a wide or radical margin was obtained, whereas after local resection the margin was intralesional in 2 cases, marginal in 8, and wide in 21. Reconstruction after resection was achieved by means of autologous or allogenous bone grafts in 13 cases, by endoprosthetic replacement in 10 cases, and arthrodeses in 2 cases.

The median follow-up period from the first operation was 8 (0.5–25) years. Of the 45 patients, histologic slides were reassessed in 33. Histologic evaluation was based on a 3-grade malignancy scale

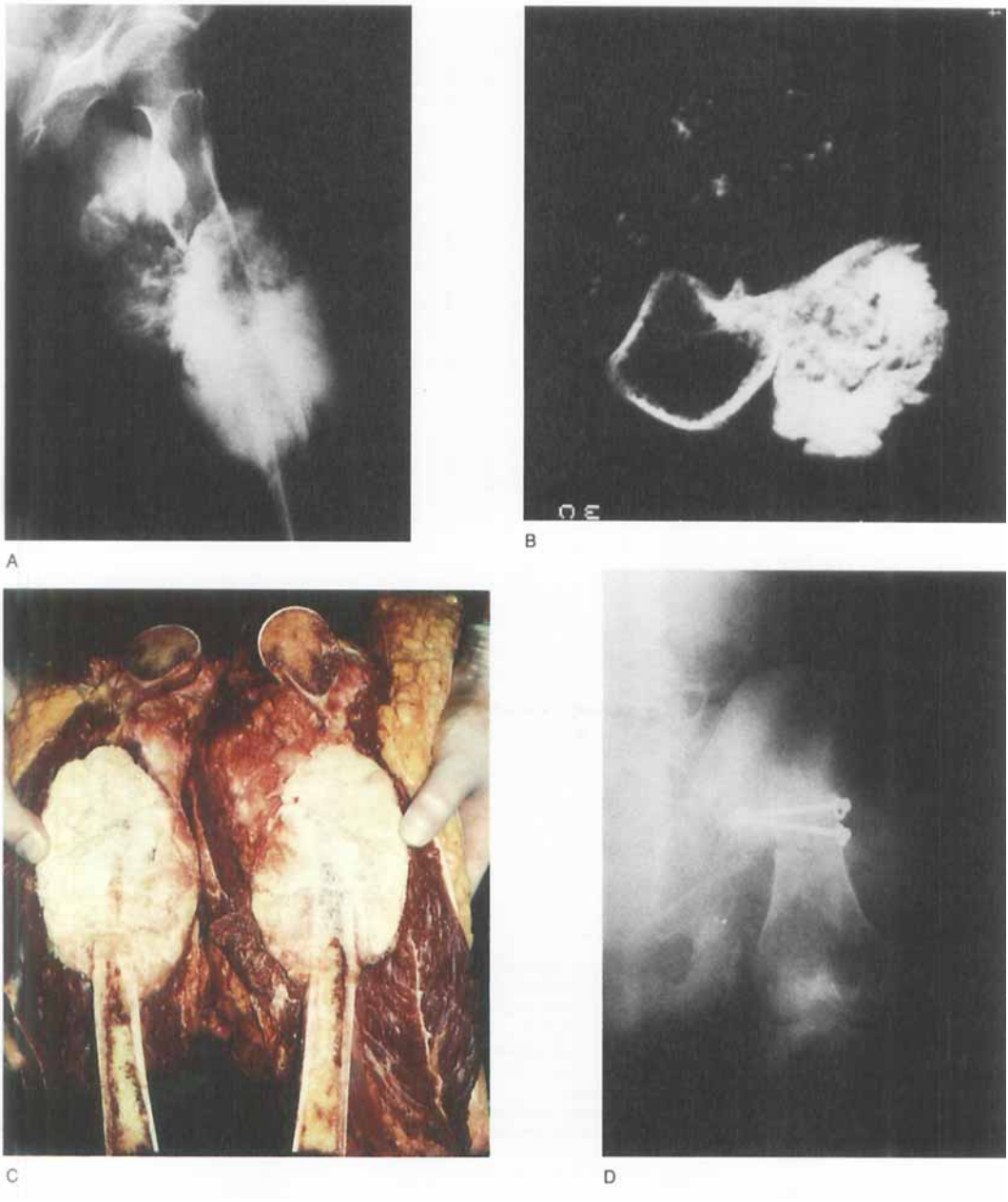


Figure 2. Case 20, 21-year-old male.

A. Parosteal osteosarcoma, Grade 1. The radiographically different mineralized areas of the tumor are conspicuous.

B. Computed tomogram of the proximal part of the tumor: gap between tumor and outer cortical bone. The intramedullary cavity shows tumor invasion.

C. Cut specimen, wide local excision. The tumor had penetrated into the medullary cavity.

D. Reconstruction was effected by means of hip rotation plasty. The function according to Enneking (1987) was poor.

according to Ahuja (1977). Evaluation of local recurrence and metastases was done only in these 33 patients (Table 2).

Results

Of the 33 histologically reassessed cases, there were 23 Grade 1 lesions, nine Grade 2 lesions, and one Grade 3 lesion.

Table 2. Data for 33 patients with parosteal osteosarcoma

Case	A	B	C	D	E	F	G	H	I	J	K	L	M	N	O	P	Q	R	S	T
1	M	f, d	26				26	2	w	1								e	4	
2	F	f, d	12	1	i	a	12	2	w	1								e	7	
3	F	f, p	59	1	i	b	60	1	m	1			6	7	r	l, b	25		6	6
4	F	f, d	16	1	(i)	c	17	2	m	1			9	5	w			f	13	
5	F	f, d	29	6	i	a	41	8	r	2						b		f	13	
6	F	t, p	30				30	2	w	2						l		g	2.5	2.5
7	M	f, d	40	1	i	d	50	7	r	1		1							15	
			40	6	i															
			49	1	i	e														
8	F	f, d	66				66	6	w	2		1				l	30			3.5
9	M	h, t	49	10		f	50	6	r	1									2	
10	M	f	17				17	7	r	2									20	
11	F	f, d	34	1	i	g	42	2	m	1		3	130	1	i				23	
													18	1	i					
													7	1	i					
													5	6	w					
12	F	r, p	72	1	i	g	73	1	m	1									7	
13	M	f, d	30				30	2	w	1	a	1						g	9	
14	F	f, d	30	1	i	g	23	2	w	1	b								8	
			32	1	i	c														
15	M	h, p	28				28	3	m	1	c	1						g	7	
16	F	f, d	30				30	4	w	1								e	2	
17	M	t, p	30				30	2	i	1									17	
							30	1	i											
							31	7	r											
18	F	f, p	16	1	i		16	4	m	1	d							g	12	
19	F	f, d	61				61	6	w	1									17	
20	M	f, p	21	10		c	21	9	w	2	e							p	3.5	
21	F	h, p	19	1	i	c	19	4	w	1								g	5	
22	M	f, d	42				42	6	w	2						l	60			7
23	M	f, d	48				48	5	w	1	f							f	5	
24	M	t, p	26				26	2	w	2								e	3.5	
25	M	f, d	39				39	9	w	2								g	2	
26	M	h	33				33	1	m	1								e	2.5	
27	F	f, d	22				22	5	w	1	g							p	5	
28	F	f	28				28	1	w	3		2							6	
29	F	f, d	16				16	3	w	1	h								5	
30	F	f, p	54				54	4	w	2									5	
31	F	h, p	26				26	4	m	1			7	7	r				6	
32	F	f, d	51				51	6	w	1									5	
33	M	f, d	39				39	6	w	1									9	

A Sex
B Site
- d distal
- f femur
- h humerus
- p proximal
- r radius
- t tibia

C Age at first operation
D Operation before referral
- 1 resection
- 2 resection + bone graft
- 3 resection + fibula interposition
- 4 resection + prosthesis
- 5 resection + arthrodesis
- 6 amputation
- 7 exarticulation
- 8 hemipelvectomy
- 9 rotation plasty
- 10 biopsy

E Margins
- i intralesional
- m marginal
- r radical
- w wide

F Histologic diagnosis at primary surgery outside tumor center
- a nonclassified sarcoma
- b inflammation
- c parosteal osteosarcoma
- d osteosarcoma
- e osteitis
- f myositis ossificans
- g osteochondroma

G Age at operation at tumor center
H Operation at tumor center, see D
I Margins, see E
J Grade
K Complications
- a pathologic fracture
- b thrombosis, amputation
- c fracture fibular graft
- d dislocation, nerve lesion
- e infection, ischemia
- f fracture
- g nail fracture
- h plate fracture

L Supportive therapy
- 1 chemotherapy
- 2 chemotherapy, COSS 82
- 3 irradiation

M Interval from first operation to local recurrence (mo)
N Kind of operation for local recurrence, see D
O Margins at operation for local recurrence, see I
P Site of metastases
- l lung
- b bone

Q Interval to metastases (months)
R Function evaluated in 16 patients (Enneking 1987)
- e excellent
- f fair
- g good
- p poor

S Follow-up (years after first operation)
T Death (years after first operation)

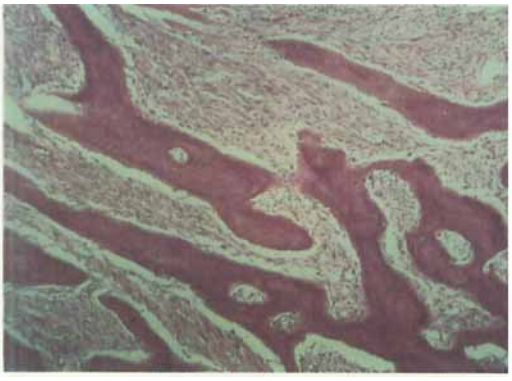


Figure 3. Case 4. Parosteal osteosarcoma—differentiated—Grade 1. Well-formed osseous trabeculae separated by a fibroblastic proliferating tissue without a significant polymorphia of the tumor cells.

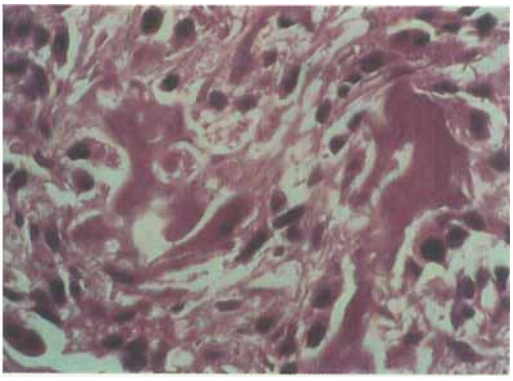


Figure 4. Case 6. Parosteal osteosarcoma—grade 2. Dedifferentiated tumor. Osseous trabeculae separated by a fibroblastic tissue with pronounced atypia of the spindle cells and malignant tumor osteoblasts.

Among the 23 patients with Grade 1 tumors (Figure 3), treatment at the tumor center comprised six amputations and 19 local resections. The margins after amputations were wide or radical, and no local recurrence occurred. The local resections were wide in 10 cases, marginal in 8 cases, and intralesional (twice) in 1 patient. After marginal or intralesional surgery, local recurrence occurred in 5/9 patients, whereas it occurred in none of the patients treated with wide resection. Pulmonary metastases developed in only 1 of the 23 patients with Grade 1 tumors (Case 3). She died 6 years after the primary operation.

Among 9 patients with a Grade 2 tumor (Figure 4), the treatment after referral was amputation in 4, local resection in 3, and rotation plasty in 2. The surgical margin was wide in 7 cases and radical in 2. There were no local recurrences. Four patients died, 3–15 years after diagnosis of pulmonary and bone metastases (Cases 5, 6, 8, and 22).

The single patient with a Grade 3 tumor received adjuvant chemotherapy according to the COSS-82-protocol. He is free of disease 5 years after diagnosis (Case 28).

Among the 25 local resections, complications occurred in 9 patients. There were 6 cases of fractures of bone grafts or implants. After prosthetic reconstruction of the hip, 1 patient had a dislocation and a transitory lesion of the femoral nerve. Finally, an amputation had to be performed after obliteration of a vascular graft (case 14).

After hip rotation plasty, 1 patient developed an infection and a soft-tissue necrosis, which was treated by excision and reconstruction with a rectus abdominus flap. Four months later, an insufficiency of the vascular anastomosis necessitated a femoro-femoral bypass (Case 20).

Discussion

We found that histologic grade was the most important prognostic factor in parosteal sarcoma, confirming previous reports (Campanacci et al. 1984, Wold et al. 1984, Dahlin and Unni 1986). Out of 23 patients with Grade 1 tumors and with one to four previous operations, only 1 patient developed metastases. In the series of Ahuja et al. (1977), 3 of 10 patients with Grade 2 tumors died of metastases. By contrast, 4 of 9 patients with Grade 2 tumors died of metastases, although all of them had had adequate surgery. Intramedullary tumor invasion has been correlated with a poor prognosis (Campanacci et al. 1984, Dahlin and Unni 1986). Our data on the pattern of tumor growth were insufficient to perform this analysis; plain radiographs do not always reveal intramedullary growth. Dedifferentiation of the tumor and a rise of the histologic grade after local occurrence have been reported (Dunham et al. 1979, Wold et al. 1984), but these were not found in our series.

The decision of whether or not to perform limb salvage (Luck et al. 1980, Kotz and Salzer 1982, Kotz 1983) or amputation depends on the individual case. Local recurrence was more common after limb salvage, but it did not seem to predispose to metastases.

Patients with Grade 2 tumors had a high incidence of metastases. These tumors should possibly be regarded as high-grade osteosarcoma. Hence, adequate treatment for patients with Grade 2 or 3 parosteal osteosarcoma may prove to be surgery and adjuvant chemotherapy.

References

Ahuja S C, Villacin A B, Smith J, Bullough P G, Huvos A G, Marcove R C. Juxtacortical (parosteal) osteogenic sarcoma: histological grading and prognosis. *J Bone Joint Surg* (Am) 1977; 59 (5): 632–47.

Campanacci M, Picci P, Gherlinzoni F, Guerra A, Bertoni F, Neff J R. Parosteal osteosarcoma. *J Bone Joint Surg* (Br) 1984; 66 (3): 313–21.

Dahlin D C, Unni K K. Parosteal osteosarcoma (juxtacortical osteosarcoma). In: *Bone tumors* (Eds. Dahlin D C, Unni K K). Thomas Cop, Springfield 1986: 308–21.

Dunham W K, Wilborn W H, Zarzour R J. A large parosteal osteosarcoma with transformation to high grade osteosarcoma: a case report. *Cancer* 1979; 44 (4): 1495–500.

Enneking W F. Modification of the system for functional evaluation of surgical management of musculoskeletal tumors. In: Enneking W F (ed). *Limb salvage in musculoskeletal oncology*. Churchill Livingstone, New York 1987; 626–39.

Geschickter C F, Copeland M M. Parosteal osteoma of bone: A new entity. *Ann Surg* 1951; 133: 790–806.

Kotz R, Salzer M. Rotation plasty for childhood osteosarcoma of the distal part of the femur. *J Bone Joint Surg* (Am) 1982; 64 (7): 959–69.

Kotz R. Possibilities and limitations of limb preserving therapy for bone tumors today. *J Cancer Res Clin Oncol* (Suppl) 1983; 106: 68–76.

Luck J V Jr, Luck J V, Schwinn C P. Parosteal osteosarcoma: a treatment oriented study. *Clin Orthop* 1980; 153: 92–105.

Salzer-Kuntschick M, Brand G, Delling G. Bestimmung des morphologische Regressionsgrades nach Chemotherapie bei malignen Knochentumoren. *Pathologie* 1983; 4: 135–41.

Scaglietti O, Calandriello B. Ossifying parosteal sarcoma. Parosteal osteoma or juxtacortical osteogenic sarcoma. *J Bone Joint Surg* (Am) 1962; 44: 635–47.

Stevens G M, Pugh D G, Dahlin D C. Roentgenographic recognition and differentiation of parosteal osteogenic sarcoma. *Amer J Roentgenol* 1957; 78: 1–12.

Wold L E, Unni K K, Beabout J W, Sim F H, Dahlin D C. Dedifferentiated parosteal osteosarcoma. *J Bone Joint Surg* (Am) 1984; 66 (1): 53–9.