Low risk of recurrence of enchondroma and low-grade chondrosarcoma in extremities

80 patients followed for 2–25 years

Henrik C F Bauer[1], Otte Brosjö[1], Andris Kreicbergs[1] and Johan Lindholm[2]

We analyzed the clinical course in 40 patients with enchondroma and 40 with low-grade chondrosarcoma of the extremities after a median follow-up of 7 years. 13 patients with enchondroma and 2 with chondrosarcoma had only open biopsy and they had no signs of further progression of the lesions. Among 23 patients with enchondroma and 23 with chondrosarcoma who were treated by intralesional curettage, 3 had local recurrences. The 10-year local recurrence rate was 0.04 in the enchondroma group and 0.09 in the chondrosarcoma group. There were no metastases. The results imply that enchondroma and low-grade chondrosarcoma of the extremities should be treated with limited surgery. The morbidity associated with en bloc resection and reconstruction can apparently be obviated without jeopardizing the limb or survival.

[1]Tumor Service, Department of Orthopedics, and [2]Department of Pathology, Karolinska Hospital, S-104 01 Stockholm, Sweden. Tel +46 8-729 2671. Fax -729 4699
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The distinction between enchondroma and low-grade intramedullary chondrosarcoma remains difficult (Meachim 1979, Smith and Monsen 1980). Both tumor types exhibit similar cellular features, such as mild atypia with occasional double nuclei (Schajowicz and McGuire 1989), and rarely mitotic figures. Pain and radiological signs of endosteal bone erosion suggest malignancy (Healey and Lane 1986). Both entities show increased uptake at scintigraphy (Hudson 1982). Quantitative DNA cytometry is rarely of diagnostic value (Kreicbergs et al. 1982).

Treatments of the two entities differ decisively. Enchondromas are either left untreated or managed with intralesional curettage (Eriksson et al. 1980). For low-grade chondrosarcomas, wide resection is generally recommended. We analyzed retrospectively our series of histologically proven enchondromas and grade I chondrosarcomas with respect to local recurrence and metastases.

Patients and methods

The study comprised 80 patients with enchondroma or intramedullary grade I chondrosarcoma of the extremities treated in our institution 1967–1991 (Table 1). Only histologically proven lesions were included (O'Neal and Ackerman 1952). Lesions of the hand were excluded, because they are known to follow a benign clinical course, almost regardless of the histological appearance (Roberts and Price 1977).

There were 40 patients with enchondroma and 40 with chondrosarcoma. 2 of the patients with enchondroma and 5 with chondrosarcoma underwent incision biopsy prior to referral; those remaining were referred with untouched lesions. None had metastases.

There was no difference in sex or age distribution between the patients with enchondroma and low-grade chondrosarcoma (Table 2). The median diameter of the lesions in the two groups was approximately the same. The majority of both types of tumors were located in the humerus or femur. The only striking difference was that there were no enchondromas in the proximal femur. Surgical staging according to Enneking et al. (1980) suggested that 20 of the 34 enchondromas were inactive. All chondrosarcomas were intraosseous, IA lesions, except 5 showing extraosseous extension, i.e., IB lesions.

The treatments of the two groups of patients were quite different (Table 3). In patients with enchondromas, one third had only an open biopsy. The remaining cases underwent intralesional curettage, except for 3 lesions of the proximal fibula (cases 16, 22, 39) and 1 of the metatarsus (case 14), resected with a marginal or wide margin (Figure 1).

Patients with grade I chondrosarcoma underwent either curettage or en bloc local resection. Two

Table 1. Data on 80 patients with low-grade chondromatous lesions

A	B	C	D	E	F	G	H	I	J	K	L	M	N
Enchondromas													
1	F	41	v	tibia, dist	3	2	1986	+	i	b		8	NED
2	M	52	v	tibia, prox	2	5	1980	+	i	b		13	NED
3	F	64	v	humerus, prox	1	3	1986	+				8	NED
4	M	16	v	radius, dist	1	4	1987	+				7	NED
5	M	64	v	femur, dist	2	1	1980	+				14	NED
6	F	51	v	femur, dist	2	3	1987	–	i	c		6	NED
7	F	52	v	humerus, prox	1	2	1988	–	i	–		6	NED
8	F	40	v	femur, dist	2	5	1987	+	i	c		6	NED
9	F	39	v	tibia, prox	1	3	1988	–	i	–		6	NED
10	M	50	v	femur, dist	2	4	1988	–	i	c		5	NED
11	F	50	v	radius, dist	1	2	1988	–	i	–		6	NED
12	F	59	v	femur, dist	2	3	1987	+	i	c		7	NED
13	F	50	v	tibia, prox	1	3	1988	–	i	–		6	NED
14	M	43	v	foot, dist	1	3	1988	–	m	–		6	NED
15	F	23	v	femur, dist	2	4	1984	+	i	b	5	1	NED
16	M	72	v	fibula, prox	1	6	1988	–	w	–		6	NED
17	F	52	v	femur, dist	2	5	1988	+				6	NED
18	M	66	v	humerus,prox	2	3	1988	+				6	NED
19	M	62	v	femur, dist	2	7	1989	–	i	b		5	NED
20	F	56	v	tibia, prox	2	3	1989	+				5	NED
21	F	39	v	femur, dist	2	2	1989	+	i	c		5	NED
22	F	53	v	fibula, prox	2	4	1978	–	w	–		16	NED
23	F	16	v	femur, dist	1	6	1984	–	i	–		10	NED
24	M	43	v	humerus, prox	1	7	1981	+	i	–		12	NED
25	F	62	v	femur, dist	1	3	1980	+	i	b		13	NED
26	F	57	v	femur, dist	2	3	1991	–	i	c		3	NED
27	F	57	v	femur, dist	1	2	1984	+				9	NED
28	F	59	b	humerus, prox	1	5	1978	+				14	NED
29	F	43	b	tibia, dist	1	7	1989	–	i	–		5	NED
30	M	45	v	femur, dist	2	10	1990	–	i	–		4	NED
31	M	61	v	femur, dist	1	6	1990	+				4	NED
32	M	54	v	femur, dist	1	6	1990	+				4	NED
33	F	71	v	tibia, prox	2	5	1990	–	i	c		4	NED
34	F	70	v	tibia, prox	1	5	1991	+				3	NED
35	F	78	v	femur, dist	2	10	1991	+				3	NED
36	F	47	v	femur, dist	2	12	1991	+	i	c		3	NED
37	F	57	v	femur, dist	1	2	1991	–	i	c		3	NED
38	F	53	v	femur, dist	1	5	1990	+				4	NED
39	F	31	v	fibula, prox	1	2	1991	–	w	–		3	NED
40	F	32	v	humerus, prox	2	3	1991	–	i	c		3	NED

A Patient number
B Sex
C Age at diagnosis
D Referral pattern
  b biopsy
  v virgin
E Location
F Surgical stage (Enneking et al. 1980)
G Largest diameter
H Year of diagnosis
I Biopsy
J Surgical margin
  i intralesional
  m marginal
  r radical
  w wide
K Type of reconstruction
  a allograft
  b bone chips
  c cement
  p prosthesis
L Time (years) to first local recurrence
M Follow-up time (years) from last procedure
N Status at last follow-up
  NED no evidence of disease
  Death, NED death without tumor

patients had open biopsy only. One, a 32-year-old man (case 49) with a lesion of the distal femur, was followed radiographically until he died of anorexia nervosa 4 years after biopsy. The other a 36-year-old woman (case 57) with a lesion of the proximal humerus, has been followed, so far, for 6 years. Among the remaining 38 patients, 23 had intralesional curettage supplemented with either autogenous bone chip transplantation or acrylic cementation (Figure 2). 8 patients underwent local resection without reconstruction. Resection and endoprosthetic replacement were performed in 9 cases, 5 of the proximal humerus, 2 of the proximal femur, and 3 of the distal femur (Figure 3). In another 2 patients, resection was followed by reconstruction with an osteoarticular allograft (case 75) or reimplantation of a resected diaphyseal segment of the femur after autoclaving (case 42). Finally, one patient (case 72) with an extraosseous lesion of a proximal phalanx of the foot underwent amputation of the toe.

Table 1. continued...

A	B	C	D	E	F	G	H	I	J	K	L	M	N
Chondrosarcomas													
41	M	45	v	tibia, dist	IB	5	1985	+	i	b		8	NED
42	M	26	v	femur, dist	IA	10	1986	+	i	a		8	NED
43	F	42	v	fibula, prox	IA	3	1986	+	m	–		7	NED
44	F	49	v	humerus, prox	IA	7	1985	+	i	b		8	NED
45	F	48	v	femur, dist	IA	6	1983	+	i	b		10	NED
46	M	65	b	foot, prox	IA	4	1980	+	i	–	3	8	NED
47	M	52	v	humerus, prox	IA	3	1982	+	w	p		11	NED
48	F	46	v	tibia, prox	IA	4	1984	+	w	b		9	NED
49	M	32	v	femur, dist	IA	12	1985	+				4	Death, NED
50	F	26	v	tibia, prox	IA	8	1985	+	i	b		7	NED
51	M	17	v	tibia, prox	IA	5	1984	+	i	b		9	NED
52	M	51	v	humerus, prox	IA	5	1983	+	i	b		7	NED
53	F	54	b	humerus, prox	IA	3	1983	+	w	p		10	NED
54	F	25	v	fibua, prox	IA	6	1987	–	i	–		6	NED
55	F	63	v	femur, dist	IA	40	1987	+	r	p		6	NED
56	M	69	b	femur, dist	IA	5	1984	+	i	c		8	NED
57	F	36	v	humerus, prox	IA	3	1987	+				6	NED
58	F	35	v	patella	IA	1	1987	–	i	b		6	NED
59	M	60	v	femur, dist	IA	3	1983	–	i	b		6	Death, NED
60	M	51	v	humerus, prox	IA	2	1980	+	w	p		13	NED
61	F	48	v	femur, dist	IA	5	1987	–	i	c		6	NED
62	F	39	v	humerus, prox	IA	3	1980	+	w	p		11	NED
63	F	37	v	femur, dist	IA	4	1988	+	i	c		5	NED
64	M	41	v	femur, dist	IA	8	1989	+	w	p		5	NED
65	M	14	v	humerus, prox	IB	4	1989	+	i	b		5	NED
66	F	69	v	humerus, prox	IA	3	1989	–	i	–		5	NED
67	F	30	v	femur, dist	IA	5	1989	+	i	c		4	NED
68	F	66	v	tibia, prox	IA	4	1988	+	i	c	1	4	NED
69	M	63	v	fibula, prox	IA	2	1979	–	w	–		14	NED
70	M	28	v	humerus, prox	IA	7	1980	+	i	b		11	NED
71	M	54	v	femur, dist	IA	3	1979	+	w	p		3	Death, NED
72	M	51	b	foot, dist	IB	2	1967	+	m		6	19	NED
73	M	36	v	femur, prox	IB	11	1984	+	i	–		8	NED
74	F	32	v	humerus, prox	IA	5	1975	+	m	p		19	NED
75	F	36	v	femur, prox	IB	10	1967	–	w	a		25	NED
76	M	69	v	femur, prox	IA	6	1990	+	i	c		4	NED
77	M	30	b	femur, dist	IA	20	1990	+	i	–		4	NED
78	F	70	v	femur, prox	IA	8	1990	+	w	p		2	Death, NED
79	M	45	v	fibula, prox	IA	3	1991	–	w	–		3	NED
80	M	18	v	femur, dist	IA	2	1991	–	i	b		3	NED

The median follow-up time for patients with enchondromas was 6 (3–16) years and for those with chondrosarcomas 7 (2–25) years. 2 patients moved abroad 6 and 7 years after diagnosis and were lost to follow-up. 4 patients (cases 49, 59, 71, 78) in the chondrosarcoma group died of unrelated causes at 2, 3, 4, and 6 years after diagnosis.

The study focused on local recurrence and metastasis. Among patients who had incision biopsy only, lesions without radiological signs of progression on follow-up were regarded as non-recurrent in the analysis.

Results

4 local recurrences occurred, 1 in the enchondroma group and 3 in the chondrosarcoma group. All recurrences developed within 6 years of diagnosis. The 10-year local recurrence rate, as assessed by the Kaplan-Meier method, was 0.04 for the enchondroma group and 0.09 for the chondrosarcoma group (p 0.3).

The single recurrent enchondroma involved a 23-year-old woman (case 15) with an intraosseous lesion of the distal femur treated with curettage and autologous bone transplantation. The lesion recurred after 6 years, was again curetted, and the cavity was filled with methacrylate cement. The tumor was again regarded as an enchondroma. She had a new local recurrence after a further 2 years and was treated in a similar fashion. The tumor was still regarded as an enchondroma, without signs of malignant transformation. There has been no recurrence 18 months after the last procedure.

2 of the 3 recurrent chondrosarcomas were extraosseous IB lesions of the foot. One of them (case 72)

Table 2. Clinical features of 80 patients with low-grade chondromatous lesions of the extremities

	Enchondroma	Chondrosarcoma
Number of patients	40	40
Sex		
male	12	21
female	28	19
Age (years)[1]	52 (16–72)	45 (14-70)
Location		
humerus, prox	6	11
radius	2	–
femur, prox	–	4
femur, dist	20	13
tibia	8	5
fibula	3	4
foot	1	2
patella	–	1
Tumor size (cm)[1]	4 (2–12)	5 (1–40)
Surgical stage[2]		
1	20	–
2	19	–
3	1	–
IA	–	35
IB	–	5
Follow-up (years)[1]	6 (3–16)	7 (2–25)

[1] Median values and range in parentheses.
[2] Enneking et al. 1980.

Table 3. Surgical treatment, margins, and reconstruction

	Enchondroma n 40	Chondrosarcoma n 40
Surgical biopsy only	13	2
Procedure		
local excision	27	37
ablative	–	1
Margin[1,2]		
intralesional	23 (1)	23 (2)
marginal	1	3 (1)
wide	3	11
radical	–	1
Reconstruction		
bone chips	5	2
acrylic cement	10	6
endoprosthesis	–	9
massive auto/allograft	–	2
none	12	8

[1] According to Enneking et al. 1980.
[2] Local recurrences in parentheses.

was a lesion of a phalanx, which recurred 6 years after a marginal toe amputation. After marginal excision of the recurrence, the patient has been free of disease for 19 years. The other (case 46) was a lesion of the calcaneus treated with curettage and autologous bone transplantation. The lesion recurred 4, 4.5, and 7 years later and was curetted each time. There has been no further recurrence since the last procedure 8 years ago. The third patient with local recurrence (case 68) had an intraosseous IA lesion of the proximal tibia treated with curettage and acrylic cementation. The patient had a recurrence after 14 months. An amputation was recommended, but the patient refused and was therefore again treated with curettage and cementation. A second local recurrence appeared after another 15 months and a thigh amputation was performed. She is free of disease 4 years later.

None of the patients has developed metastases, nor has anyone died of tumor-related causes during the follow-up period.

Figure 1. Case 8.

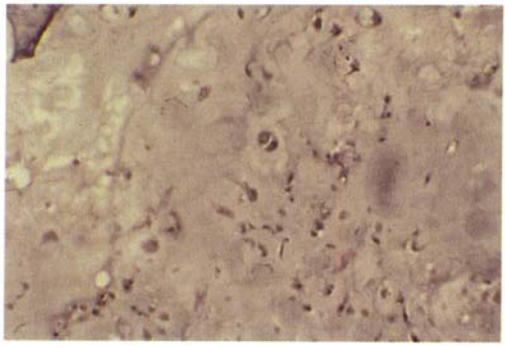
Enchondroma of distal femur in a 40-year-old woman.

Histology showed largely acellular cartilagenous material.

After curettage and cementation. No recurrence after 6 years.

Chondrosarcoma grade I of distal femur in a 69-year-old man. Note lytic area around mineralized tumor.

Figure 2. Case 56.



Histologic appearance showing increased cellularity.

7 years after curettage and cementation.

Figure 3. Case 74.

Chondrosarcoma grade I of proximal humerus in a 32-year-old woman. Note medial cortical erosion. Treated 1975 with wide resection and reconstruction with an isoelastic prosthesis.

Follow-up at 9 years showed cranial dislocation resulting in poor function. Today we would have performed curettage and cementation of the lesion.

Discussion

Our study shows that local recurrence is clearly rare in enchondroma and also in low-grade chondrosarcoma, even after non-radical surgery.

The low rate of local recurrence would seem to imply that our classification of chondrosarcoma grade I was too lenient; others, indeed, might have classified them as enchondromas. However, the lesions were relatively large, the majority were located centrally in the humerus or femur, and the patients had a clinical history of pain. Commonly, there were also radiologic signs of endosteal activity. Hence, we believe that the classification by and large was correct.

The observation that only 1 of 40 enchondromas recurred suggests that it is not necessary to treat these patients, even the necessity of open biopsy appears questionable. On the basis of plain radiography, it is usually possible to establish whether the lesion is an enchondroma or a low-grade chondrosarcoma (Eriksson et al. 1980). The histologies of these two entities are similar, and thus a biopsy is hardly helpful. In our opinion, if there is no pain and the lesion is radiologically inactive, a biopsy is not necessary. However, the patient should be asked to return should clinical symptoms arise.

For painful lesions with radiological signs of activity, the diagnosis of chondrosarcoma is more likely. Open biopsy, followed by local resection and, if necessary, reconstruction is recommended (Eriksson et al. 1980). As the morbidity of major skeletal reconstructions is considerable, the procedures should be confined to tumors with a considerable risk of local recurrence or metastasis. Our findings indicate that this is not the case with central grade I chondrosarcoma of the extremities. In our series, 17 patients with lesions of the femur or humerus had inadequate margins, after either intralesional curettage or marginal

resection. Yet, none of these lesions recurred. Hence, we believe that a central grade I chondrosarcoma of long bones can be treated with curettage and filling the cavity with either autogenous bone or methylmethacrylate cement. Distal destructive lesions require en bloc resection to prevent local recurrence. Chondromatous lesions of the pelvis and shoulder girdle must be treated aggressively, since they are known to be associated with a high risk of local recurrence, which is even more difficult to control (Sanerkin and Gallagher 1979).

The fact that none of the patients with low-grade chondrosarcoma developed metastases throws doubt on malignant nature of this entity. In the differential diagnosis between enchondroma and grade I chondrosarcoma of the extremities, the former diagnosis, in our opinion, should be preferred, unless there are convincing clinical and radiological signs of malignancy.

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