

CLINICAL STUDY

Parosteal osteosarcoma

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Abstract: In this retrospective clinical study, 6 cases of osteosarcoma of the bone have been analyzed. Five patients were with parosteal osteosarcoma and one with periosteal osteosarcoma. The study was performed at the Clinic for Orthopaedic Surgery in Skopje, Macedonia, from 1995 to 2005. This tumor represents 1.5 % of all primary bone tumors treated at the Clinic in the 11 year period. The age of the 6 patients (2 female and 4 male) ranged from 8 to 39 years (average 23.8). The history analysis of the patients showed misinterpreted diagnosis in 50 % of the cases, with 83.3 % rate of local recurrence, 33.3 % of metastases and 33.3 % of mortality. Follow-up varied from 11 months to 9 years (average 4.5). The clinical and histopathological findings (identical with those reviewed in the literature) confirmed occurrence of two biologically different types of parosteal osteosarcoma: predominant type is originally “benign” but has a definite malignant potential, causing metastases after long symptom-free interval. The other type is highly malignant from the beginning. More radical surgery is recommended for the latter category of tumors, followed by chemotherapy. Compartmental, radical “en bloc” resection, followed by regular review of the patients, is recommended for the former (Tab. 1, Fig. 3, Ref. 20). Full Text (Free, PDF) www.bmj.sk.

Key words: parosteal osteosarcoma.

Parosteal osteosarcoma is a rare low-grade bone tumour. It was apparently described for the first time in 1951 by Geschickter and Copeland, regarding the initial confusion with the terminology (6, 7, 21). It occurs between the 2nd and 7th decade of life and it represents 1.6 to 2 % of all malignant bone tumours (13). The most frequent location is the distal dorsal femur. Until clearly proven otherwise, a bone-forming tumor in this localisation must be regarded as parosteal osteosarcoma. To determine the histopathological diagnosis could be “tricky”. The tumor is characterised by hyalinized fibrous stroma with low cell content without substantial nucleus polymorphism and variably dense bony trabeculae. The operation material must undergo a careful pathohistological analysis, because the extent of invasion of the medullar cavity and most probably the extent of dedifferentiated areas determines the prognosis and occurrence of local recurrence and metastases (3, 4).

As most authors report, a wide margin of excision ensures adequate surgical treatment of parosteal osteosarcoma in any surgical grade or stage. No evidence for the development of primary tumor satellite nodules or of “skip” metastases were seen, so it would seem that truly radical or compartmental surgery is rarely indicated. The significant incidence of pulmonary me-

tastases among those patients with Grade III parosteal osteosarcoma and involvement of the medullar cavity, suggests that, for them, adjuvant chemotherapy should be considered (1, 2, 3, 13). Primary wide excision may be less effective for local recurrence where there has been a previous inadequate biopsy or surgical treatment, because of contamination and spread of the tumor into the surrounding tissues (2).

The tumor is most commonly misinterpreted as osteochondroma or heterotrophic ossification and even large institutions have limited experience of its diagnosis and management (3). Parosteal osteosarcoma shows like no other tumor the necessity of close cooperation of all involved disciplines for diagnosis and therapy and should be treated only in specialized institutions for bone tumor surgery.

Patients and methods

At the re-examination of the records at the Clinic for Orthopaedic Surgery in Skopje, during the last 11 years (from 1995 to 2005), 6 cases of juxtacortical osteosarcoma were found. Five of them were patients with parosteal osteosarcoma and one patient was with periosteal osteosarcoma. Parosteal osteosarcoma represented 1.5 % of all 396 patients with malignant bone tumors treated in this time at the Clinic.

Reviewing the records, bone scans with Technetium 99m, radiographs, arteriography, CT, MRI and histopathology findings showed 5 patients with confirmed diagnosis of parosteal osteosarcoma and one was with high grade chondroblastic type of periosteal osteosarcoma. Four of them were diagnosed and treated at the Clinic for Orthopedic Surgery in Skopje and two

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Tab. 1. Clinical details of the 6 cases.

Case/ /initials	Sex	Age	Site of the tumor	Duration of symptoms (months)	Symptoms	Treatment	Chemotherapy	Local recurrence	Metastases	Follow-up (months)	Outcome (MSTSS)
1. VM	F	28	proximal tibia	13	painless tumor	1. radical resection +MMA	after oper.	after 6 months	no	23	25
2. BT	M	27	distal femur	15	blunt pain and tumor	1. resection 2. reresection 3. amputation	after oper.	after 3 months	lungs after 23 months	37	30†
3. BS	M	8	distal femur	9	blunt pain and tumor	1. resection arthrodesis 2. recurrence excision	neo-adjuvant	after 3 months	lungs after 16 months	21	37†
4. SV	F	39	distal femur	17	painless tumor	1. resection arthrodesis 2. ste.	after oper.	after 32 months	no	71	39
5. ID	M	24	proximal femur	13	painless tumor	1. resection and ste. 2. disarticulation after masive local recurrence	neo- adjuvant	after 61 months	no	67	31
6. JM	M	17	proximal humerus	8	severe pain tumor and reduction of movements	1. radical resection + nonvascular fibular graft	no	no	no	109	33
			average 23.8	average 12.5			average 37	19.5	54.7	32.5	

Legend: MSTSS – Musculoskeletal Tumor Society Score (5), MMA – reconstruction of the defect with metilmetacrilate (bone cement), Resection arthrodesis – reconstruction with intramedular nail and MMA, STE – special tumor endoprosthesis, † – lethal outcome

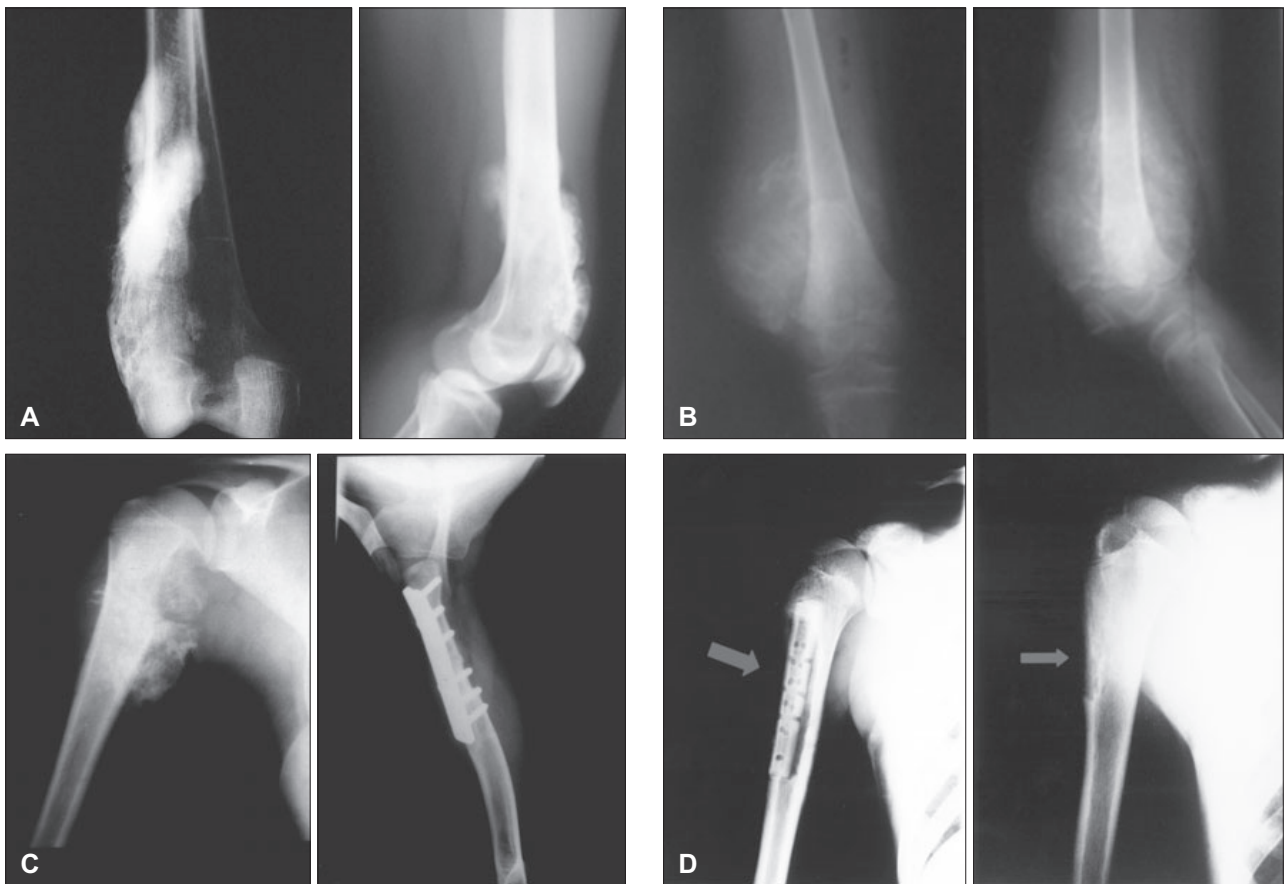


Fig. 1. a) frontal and lateral radiography of the parosteal osteosarcoma of right distal femur (case 2); b) frontal and lateral radiography of local recurrence of “high grade” chondroblastic type periosteal osteosarcoma (case 3); c, d) densely ossified and lobulated parosteal osteosarcoma of the right proximal humerus, the tumor was radically resected and the bone was reconstructed with fibular graft (case 6).

patients started their treatment elsewhere and were misdiagnosed for osteochondroma. Another three started their treatment as unspecified malignant tumor of distal femur and only one was primarily suspected to be parosteal osteosarcoma.

Results

The clinical details of all 6 patients are summarized in Table 1.

Age and sex. The age ranged from 8 to 39 years (average 23.8). There were 2 female and 4 male patients.

Site of the tumor. In 3 cases the site of tumor was the posterior surface of the distal femur. In 1 case primary site of tumor was postero-medial surface of the proximal femur. In 1 case the site of tumor was proximal humerus and in 1 case the site of tumor was the posterior surface of the proximal tibia.

Symptoms. The most frequent sign on admission at the Clinic was localized painless swelling present in 3 patients, progressively increasing blunt pain and swelling was present in 2 patients and severe “night pain”, swelling and restriction of movements was present in 1 patient. Duration of the symptoms varied from 8 to 17 months (average 12.5).

Radiological findings. In 3 cases the radiographs showed a densely ossified and lobulated mass on the posterior metaphyseal cortex of the distal femur (Fig. 1a). Similar dense and lobulated tumors were seen at the other sites (Figs 1c, 1d). Tumors were attached to the bone by broad base. In one case, after the recurrence, two thirds of the circumference of the distal femoral metaphysis were involved (Fig. 1b). This patient was considered to have medullar involvement on CT scans.

Pathological findings. The tumors were ossified with occasional soft areas, all of them infiltrating the surrounding soft tissues. Most of them had typical histological appearance of a low grade parosteal osteosarcoma, with spindle cells and collagen fibers embedding osseous trabeculae. The spindle cell stroma was scarcely cellular, with low to moderate atypia of the cells, as well as low mitotic activity. The trabeculae were rather regularly arranged, but missing the osteoblastic rimming. Two of the cases also showed cartilaginous islands in the mainly fibroblastic tumour tissue (Fig. 2a). Most of the cases had well (G1) to moderate (G2) degree of differentiation (Fig. 2b).

Only one presented a high grade chondroblastic (G3) surface osteosarcoma. It was presented as a high grade surface chondroblastic osteosarcoma (case 3) (Tab. 1), with scarce and hard to find foci of osteoid and wide areas of malignant cartilage. The tumor cells showed marked polymorphism and high mitotic index (Fig. 2c). Recurrent lesions usually showed less differentiation than the primary tumors.

Medullar involvement. In the 6 patients managed, 1 had initial histological medullar involvement and 2 patients had medullar involvement after the local recurrence (Fig. 3a). Two patients who were treated out of the Clinic for Orthopaedic Surgery in Skopje initially, it was not possible to tell, due to lack of evidence (Figs 3b, 3c). Only one patient had no medullar involvement (case 6).

Discussion

Parosteal osteosarcoma is a rare malignant bone tumor first described by Geschickter and Copeland in 1951 (6, 7, 12, 19). Up to date, there are reports of parosteal osteosarcoma even in human pets (20). Larsson and Lorentson found only 206 cases (including sixteen cases from their study) to be reported until 1980 in the world literature. The annual incidence in Sweden, as they reported, corresponds to one case per 8 000 000 inhabitants and were accounted for about 2 % of all primary malignant tumors of bone and 6.2 % of all osteosarcomata. In comparison, Dahlin reported that parosteal osteosarcoma constituted only 3.7 % of all osteosarcomata from Mayo Clinic (9, 13, 17).

Most of the world studies documented difficulties in the diagnosis of parosteal osteosarcoma. The inability to diagnose the lesion correctly often leads to inadequate initial operative procedures. The differential diagnosis may include diverse entities such as: myositis ossificans, fracture callus, ossifying haematoma, osteochondroma, extraosseous osteosarcoma, parosteal chondroma, desmoplastic fibroma and osteoma (2, 3, 4, 13, 16).

The clinical characteristic of patients who have a parosteal osteosarcoma is distinctly different from that of patients who have conventional osteosarcoma (7, 16). The most common complaint was “painless swelling”, same as reviewed in the literature, presented with 3 patients (2, 3, 12, 13, 17). Two of our

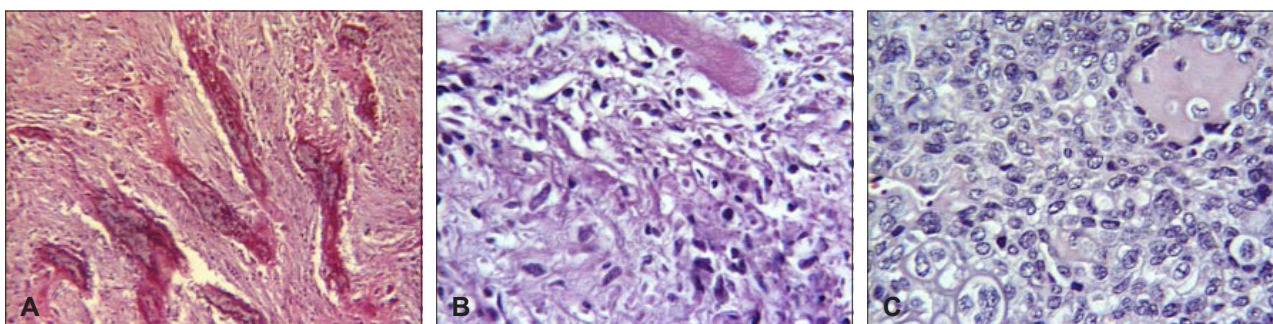


Fig. 2. a) parosteal osteosarcoma showing parallel osteoid trabeculae embedded in fibroblastic stroma. HE, x100, b) focus of a moderate grade of differentiation (G2) in a well differentiated osteosarcoma. HE, x200, c) Osteoid deposits in high grade chondroblastic osteosarcoma. HE, x400.

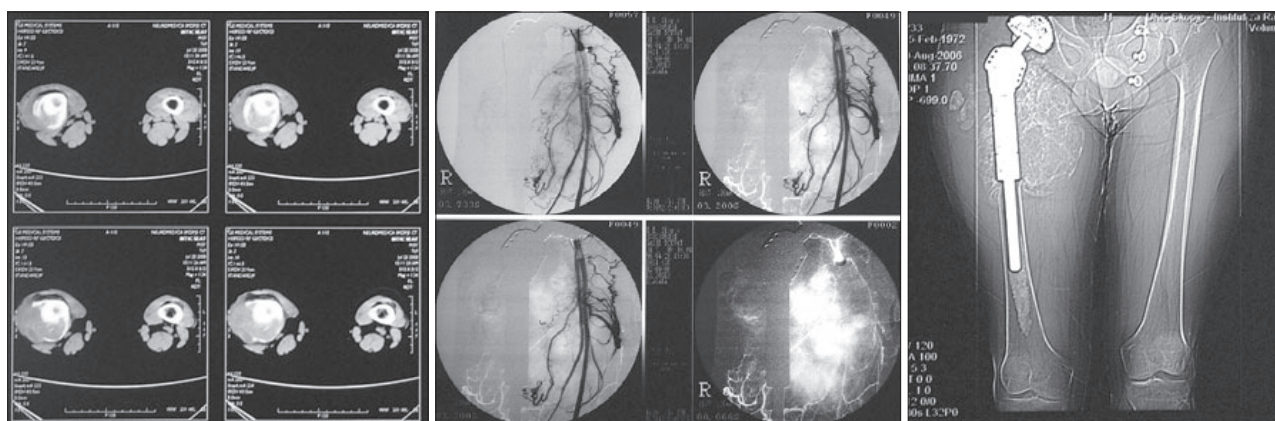


Fig. 3. a) CT of distal femur showing the extent of medullar involvement of the bone (case 3), **b)** arteriography of the proximal right thigh showing special tumor endoprosthesis and pathological vascularisation due to local recurrence (case 5), **c)** CT of the recurrence of parosteal osteosarcoma at the proximal femur after limb salvage operation with special modular endoprosthesis (case 5).

patients had “blunt pain with swelling” and one had “night pain”. Most of our patients had symptoms of prolonged duration more than one year before admission at the Clinic.

The site of the parosteal osteosarcoma in our study correlates with the reported sites in the literature. A predilection for anatomic site was characteristic feature of the patients in the study and showed that 50 % of the patients in our study had involvement of the posterior part of distal metaphysis of the femur (2, 3, 14, 15, 19).

The well described concept of dedifferentiated parosteal osteosarcoma with higher incidence of development of metastases is also applicable in our study (4, 8, 13, 18). Dedifferentiation of the tumor showed high incidence with pulmonary metastases in one of our patients. Long-term follow up is essential for assessing the real malignant potential of parosteal osteosarcoma since recurrence can be considerably delayed (10).

The results of various types of therapy are always difficult to evaluate in retrospective manner. A review of the literature shows that local excision of the tumors has almost invariably resulted in recurrence (1, 10, 13, 16, 18, 19). Local recurrence was not related to medullar involvement of the tumor but to the number or adequacy of biopsies and surgical margins of the resection of the tumor (11, 14). Two of the patients had more than one, and one patient had three inadequate placed biopsies. Further more three of the patients in our study had intralesional or inadequate marginal resection of the tumor during the primary surgical treatment. Overall, results were poor to fair and varied from 25 to 39 points of MSTS score (5).

Conclusion

The findings of this study followed the theory that two distinct types of parosteal osteosarcoma exist: one type, which is primarily highly malignant and the other one, which is originally benign but with inherent malignant potential. Only two patients of our study had modern chemotherapy and this did not allow any definite conclusion. In most of the cases the histo-

pathological diagnosis was a problem at the beginning of the treatment. For well delineated tumors with well-differentiated histological appearance, radical (en block) resection of the tumor and surrounding soft tissue is strongly recommended. In patients with advanced tumors containing pleomorphic areas and/or inadequate placed biopsies or prior inadequate surgical treatment, amputation should be undertaken. Parosteal osteosarcoma shows like no other tumor the necessity of close cooperation of all involved disciplines for diagnosis and therapy and should be treated only in specialized institutions for bone tumor surgery.

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