Periosteal Osteosarcoma
A Case report

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Abstrak

Periosteal osteosarkoma merupakan tumor ganas tulang yang jarang didapat, dibentuk dari sarkoma tulang dengan didominasi komponen tulang rawan yang berdiferensiasi dan tumbuh pada permukaan tulang. Penelusuran kepustakaan tidak banyak menyebutkan mengenai kasus ini. Laporan kasus ini terakhir dilaporkan oleh Klinik Mayo tahun 1999. Kami laporkan satu kasus periosteal osteosarkoma pada penderita laki-laki berusia 17 tahun. Penderita menjalani tindakan pembedahan berupa prosedur "limb salvage", dengan pra dan pasca bedah penderita mendapat kemoterapi (neo-ajuvan dan ajuvan). Tidak ditemukan rekurensi lokal dan metastasis di paru, pada follow up sampai dengan 14 bulan pasca bedah. ***(Med J Indones 2003; 12: 166-70)***

Abstract

Periosteal osteosarcoma is a rare type of malignant bone neoplasm, with predominantly cartilaginous component and arising on the bone surface. Reports of the case in the literature were rare. Last case was reported by Mayo Clinic in 1999. We report a case of periosteal osteosarcoma in a 17-year-old male, who was treated surgically with a limb salvage procedure, neoadjuvant and adjuvant chemotherapy were also given to the patient. There was no local recurrence and lung metastases up to 14 months after surgery. ***(Med J Indones 2003; 12: 166-70)***

Keywords: *osteosarcoma, periosteal osteosarcoma, limb salvage.*

Periosteal Osteosarcoma is a rare subtype of osteosarcoma, which is one of other surface osteosarcomas besides parosteal osteosarcoma and high grade surface osteosarcoma. Periosteal osteosarcoma is a low to intermediate grade bone forming sarcoma with predominantly chondroblastic matrix as the main component. This tumor arises from the deeper layer of the periosteum on the surface of bone, especially in the long bones with elevation of the periosteum and prominent periosteal new bone formation.[1,2]

Periosteal osteosarcoma represents 1-2% of all osteosarcoma and accounts for 25 % of juxtacortical osteosarcomas.[1,2,3] Although the ages of the patients vary, mostly occur in the second and third decades of life and the tumor occurs more commonly in female patients with a 1:1,7 male/female ratio.[1,2] The overall prognosis of periosteal osteosarcoma is somewhat better than that conventional osteosarcoma, but worse than parosteal osteosarcoma.[2]

Periosteal osteosarcoma was first identified as a distinct type of tumor by Unni et al in 1976.[4] The last study was reported by Mayo Clinic in 1999, from 1649 osteosarcoma cases, 26 cases were periosteal osteosarcoma (1,5%).[2] Based on Department of Anatomic Pathology, Faculty of Medicine University of Indonesia data from January 1995 till January 2002, there were 238 bone tumor cases, among them there were 83 cases osteosarcoma (34,87%) and only one case periosteal osteosarcoma (1,2%).

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CASE REPORT

A 17-year-old male was referred to the Cipto Mangunkusumo Hospital from Riau, Central Sumatra for further evaluation of possible malignant bone tumor on his right distal diaphysis of the tibia. He had a five months history of the lump and pain on his right leg. The result of the biopsy in Riau was conventional osteosarcoma (intramedullary) type.

Physical examination showed a lump on the right distal diaphysis of the tibia in anteromedial portion. Its dimension was 7,5x5x1 cm, hard, fixed and a biopsy scar was visible on the surface of the mass. A complete blood cell count and blood chemistry examination were within normal limits.

Plain radiographs of the right tibia showed irregularity and thickening on the surface of the cortex with spiculated and Codman triangle periosteal reactions. Magnetic Resonance Imaging (MRI) showed a mass on the right distal third tibia with erosion of the cortex and no involvement of the medullary cavity.

A clinical pathological conference was conducted between the orthopedic oncologist, radiologist, pathologist, and medical oncologist. The meeting concluded that the tumor was a periosteal osteosarcoma on the right distal third of the tibia. Based on that diagnosis the patient was planned to have surgery with neoadjuvant chemotherapy before hand.

The patient underwent a three cycle neoadjuvant chemotherapy with cisplatin and adriamycin with three weeks interval. After neoadjuvant chemotherapy the pain was gone.

After 3 cycles of preoperative chemotherapy, surgery was performed with limb salvage procedure. Resection was done 5 cm proximal to the mass on the tibia and distally down to the ankle joint. The resected bone segment underwent 15 minutes autoclaving process, and then re-implanted. Fixation was done with plate and screw. Arthrodesis was performed in the ankle joint. Before the autoclaving process, the mass on the bone segment was taken for histologic specimen evaluation and the result was less than 50 percent necrosis. (Huvos I).

After that a second clinical pathological conference was performed and because the result of preoperative chemotherapy was less than 50% tumor necrosis, we decided to change the regimen of chemotherapy. The new regimen consisted of cisplatin, doxorubicin, high dose methotrexate, bleomycin, ifosfamide and dactinomycin that would be given in three cycles.

At his latest follow-up visit 14 months post operatively, the patient remained disease free with no evidence of metastasis. Plain radiograph of his right leg showed no local recurrence and loosening of the screws.

DISCUSSION

Osteosarcoma is the most common malignant tumor of bone. It covers a wide spectrum of lesions with distinct clinical and pathological features, associated with different biological behavior. According to the WHO 1993 classification, osteosarcoma can be separated in the two fundamental groups, the central (intramedullary) and surface osteosarcoma.[5] Periosteal osteosarcoma is a subtype of the surface osteosarcoma beside parosteal osteosarcoma, and high grade surface osteosarcoma.[5] This tumor was firstly reported by Unni et al in 1976, incidence rate is 1-2% of all osteosarcoma and 25% of all juxtacortical osteosarcomas.[2,3]

Literature review showed that this tumor occurs more commonly in female with a 1:1,7 female ratio.[1,2] Periosteal osteosarcoma in this case occured in a young man, 17-year-old and based on the age is in accordance to the literature that this tumor occurs commonly in the second and third decade.[1,2]

In our case the tumor occurred in distal third of the diaphysial tibia, and this is in accordance to the literature review that tumor occurs mostly in the long tubular bones of the lower extremity.[1,2] Literature review showed that the tumor occurs pre dominantly in the diaphysis as our case does.[1,5] As we found in our patient, the literature review showed that the tumor mostly is located on the anteromedial portion of the tibia.[6]

Periosteal osteosarcoma is a predominantly radio lucent fusiform lesion with elevation of the periosteum that presents on the surface of a long bone.[1] Radiographic characteristics were defined by deSantos et al[7] and include nonhomogeneous tumor matrix spiculated interspersed with an area of radiolucency, representing uncalcified matrix, occasionally a periosteal reaction in the form of

Codman's triangle, thickening, irregularity and erosion of the cortex without medullary involvement.[2] Plain radiographic studies in our patient showed thickening and irregularity of the cortex with spiculated and Codman triangle forms of periosteal reaction.

Some authors use the absence of medullary involvement as a *conditio sine qua non* for diagnosis of this tumor. MRI can evaluate the involvement of the medullary cavity.[1,2] MRI evaluation in our patient showed no medullary cavity involvement.

Based on clinical data, radiologic and histopatologic examination, clinicopathological conference was performed and the conclusion was periosteal osteosarcoma.

The patient then was planned to have limb salvage procedure and neoadjuvant chemotherapy. Cisplatin and adriamycin were given as neoadjuvant chemotherapy as much as three cycles with three weeks interval. Literature review showed that the aims of neoadjuvant chemotherapy is reduction of tumor size, increasing the chance of limb salvage, reduced drug resistance possibility, provide time for endoprosthesis fabrication, less chance of viable tumor being spread at the time of surgery and potentially increases surgical outcomes.[8,9] Today neoadjuvant chemotherapy has become a part of standard osteosarcoma treatment.[9]

The treatment of an osteosarcoma patient is surgery and adjuvant chemotherapy. Surgical treatment could be limb salvage or limb ablation.[10,11] Thirty years ago, a high grade bone tumor patient would had amputation as local control.[12] Today along with the development of multiagent neoadjuvant chemotherapy, the treatment of osteosarcoma is limb salvage and reconstruction.[11,12]

Retrospective study between osteosarcoma which underwent amputation and limb salvage procedure showed that there was no significant differences in local recurrence between amputation (5%) and limb salvage (5-10%), no differences in survival (70%), and limb salvage offer better result in functional outcome and long term cost. Today 85-90% osteosarcomas in the extremity underwent limb salvage procedure.[12] In limb salvage procedure, after resection of the tumor the defect is reconstructed using either allograft, autograft or metallic prostheses.[13]

Limb salvage procedure was done in our patient with resection and reconstruction by using autoclaved-autograft. Ideally limb salvage procedure is using allograft, metallic prostheses or alloprosthetic composite. Limb salvage using autoclaved-autograft technique was chosen because bone bank services is not yet available.[14] This alternative reconstruction technique firstly was introduced by Steggal and Thompson in 1956.[15]

The recent report about reconstruction in using autoclaved-autograft technique for bone tumor was reported in Germany in 1997 for 9 cases of lower extremity bone tumor and in Singapore during 1980-1999 for reconstruction of 14 osteosarcoma cases in the lower extremity.[16,17]

Schajowics et al described the prognosis of his nine periosteal osteosarcoma patients, who underwent wide resection. Three of his nine patients died caused of lung metastasis.[2] The rate of metastasis in periosteal osteosarcoma was 15-20% mostly in the lung, because of that, in periosteal osteosarcoma it is suggested to give adjuvant chemotherapy to eradicate the metastatic foci.[1,2,9,11]

Extent of the disease, primary site of the tumor, skip metastasis, pathologic fracture, and histologic evaluation of the percent of tumor necrosis are prognostic factors of osteosarcoma.[18] In our case, we found unfavorable prognostic factor, that was necrosis of the tumor which showed Huvos I. This unfavorable result could be "salvaged" by adding a new agent in post operative chemotherapy.[18]

Clinically the result of neoadjuvant chemotherapy showed decrease in pain, that was a good clinical prognostic factor.[11] Initial high level of alkaline phosphatase reveal unfavorable factor.[11] In our patient, we did not find initial elevation level of this enzyme and also up to the last 10 months follow up.

Overall, it appears that periosteal osteosarcoma metastasize in 15-20%, and mostly in the lung.[1,2] Our patient was controlled regularly, and after 14 months postoperative we did not find lung metastasis. X-ray evaluation of the right leg after fourteen months post operative didn't showed local recurrence, there was callus formation on the site of the osteotomy and distally the ankle joint was mostly fused and patient was told to walk full weight bearing with the aid of one crutch. The patient is still under our observation to detect any local recurrence and distant metastasis.

Figure 1. Preoperative AP and lateral plain radiograph showed mass on the surface of tibial cortex with spiculated and codman triangle periosteal reaction

Figure 2. MRI of the patient showed the tumor on the surface of tibia that had eroded the cortex and no involvement of the medulla

Figure 3. Fourty months post operative AP and lateral plain radiograph, showed callus formation on the side of osteotomy; arthrodesis of the ankle joint was mostly achieve

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