Neglected Osteosarcoma of the Knee in a Young Female Adolescent: A Case Report.

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ABSTRACT

A 16-years old girl presented to the orthopaedic outpatient clinic of Dr. Zainoel Abidin Hospital, Banda Aceh, Indonesia with a progressive growing mass around the left knee since 12 months ago. Diagnostic assesment includes clinical, radiological and fine needle aspiration cytology of the mass confirmed an osteosarcoma. Patients undergo above knee amputation procedure as definitive treatment. Neglected osteosarcoma is a primary malignant bone tumor with more than 3 months delayed from the original time of signs and symptoms occurrence to first presentation for medical care. The combination of unawareness, low socio-economic level and inability to access healthcare facilities are major factors contributing to delayed presentation in this case. Management of neglected osteosarcoma in our region faced a variety of issues such as lack of funding, inadequate diagnostic facilities and limited access to advanced treatment options combined with patient's cultural and socio-economic preferences with regards to management and treatment options.

Keywords: Amputation, Health services accessibility, Osteosarcoma, neglected, Disease management.

INTRODUCTION

Osteosarcoma (OS) is the most common primary malignant bone tumor, usually occurring in the second decade of life.[1] The management of patients with osteosarcoma in developing countries particularly in our geographical region is challenging and often delayed due to the lack of awareness amongst patients of their illness, patients' cultural preferences and poor socio-economic status, coupled with inadequate diagnostic facilities and effective therapy issues.[2] These factors often led to delayed presentation of malignant bone and soft tissue tumours to medical care, more recently labeled as neglected malignant bone tumours, when presentation to medical care is delayed by more than 3 months from time of signs and symptoms.[3,4] Neglected malignant bone tumours because of their advance stage tend to be associated with a poorer prognosis with high morbidity such as limb loss and mortality.[3] Clinical presentation such as severe pain, tumor fungation, sepsis, thrombosis, hemorrhage, pathologic fractures and severe functional impairment is often the case. We present here a case of neglected advanced osteosarcoma of the left knee in a young teenage girl and discussed the management options available.

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CASE REPORT

A 16-years old girl presented to the Orthopae-

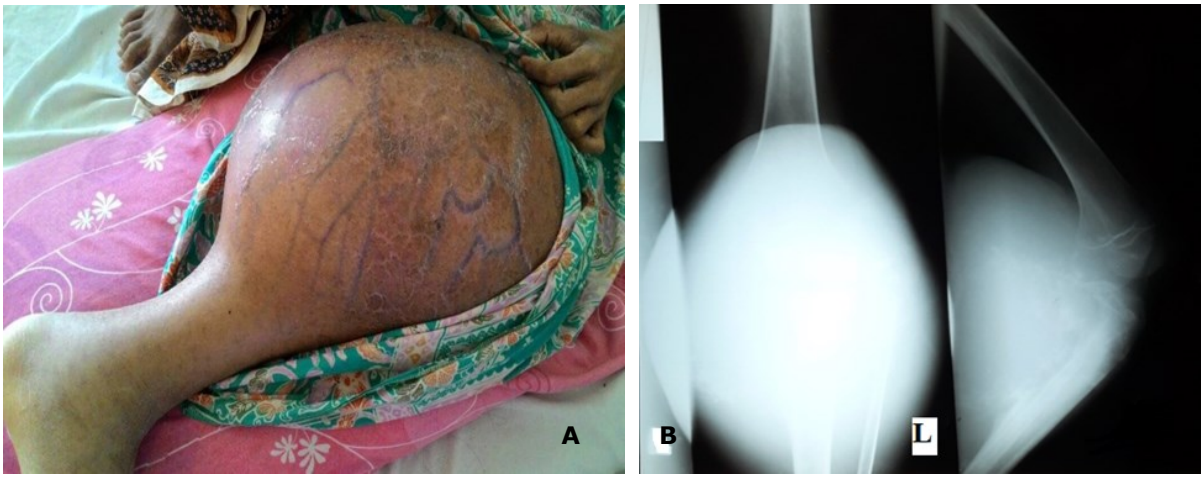

Figure 1. A) Mass around the left knee joint; B) Plain x-ray of the mass (Left Knee Joint)

dic Outpatient clinic of Dr. Zainoel Abidin Hospital, Banda Aceh, Indonesia with a progressively enlarging tumour around the left knee of 12 months duration. The patient suffered from severe pain in the knee with inability to walk normally since 7 months ago. She gave a history of previous left lower leg trauma a year ago which was treated by a traditional bone-setter using massage and traditional herbal medicine around the injured knee.

Physical examination showed poor general condition with underweight appearance. Vital signs were within normal limit. There was a large solid tumour measuring 39 cm by 32 cm, arising from lower third of the thigh and extending across the left knee to mid shin. The skin overlying the mass was thin and ulcerated in some areas with underlying dilated veins (Figure 1a). The range of motion of left knee was severely limited.

Blood investigations showed mild anemia with Hb of 9.0 mg/dL and an elevated white cell Count of 18.3 x 10^3mm^3 and platelets of 646 x 10^3U/L. Plain radiograph of the left knee showed well-defined soft tissue tumour arising from the upper third of the left tibia with expansile lytic lesions, periosteal reaction (Codman triangle), sunray appearance, lateral shifting of tibia-fibula bone and extending upwards to the lower third of the femur and downwards to the mid shin level (Figure 1B). Fine needle aspiration cytology of the tumour showed clusters of osteocytes with hyperchromatic pleomorphic nuclei, increased Nuclear-Cytoplasmic (NC) ratio and producing osteoid (Figure 2).

Given the advanced stage of disease, a plain chest radiograph and ultrasonography of abdominal organs were performed for the initial staging of her disease. The chest radiograph was clear and ultrasonography showed no liver or intraabdominal metastatic lesion. She underwent above knee amputation as a definitive treatment for the advance osteosarcoma. She was followed up postoperatively after her discharge up to a year where she was well but had refused to undergo postoperative CT staging or adjuvant chemotherapy.

DISCUSSION

Osteosarcoma is primary mesenchymal malignancy characterized by the production of osteoid.[1] The incidence of Osteosarcoma showed a bimodal peak in life, the first peak occurs in the adolescent and young adult and a second peak in the 6th decade of life.[2] Even though, there have been significant improvements in the diagnosis and treatment of osteosarcoma,

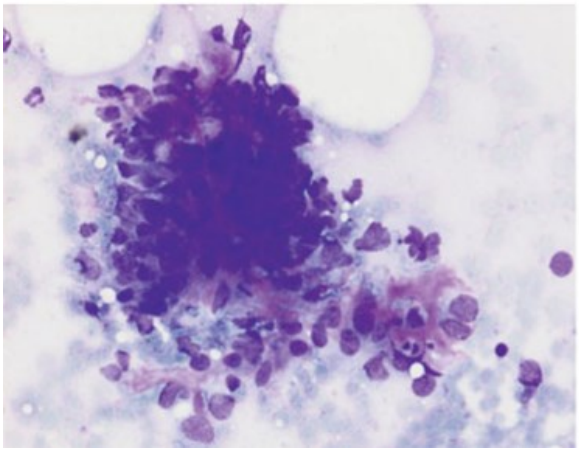

Figure 2. Fine Needle Aspiration smears of Osteosarcoma (MGG stain, original magnification x25)

overall survival rates are relatively constant for the past 2 decades. The extreme variability of osteosarcoma cases from one to another, making single treatment pathway seems unlikely to be able to treat all or even most of the cases.[2]

The occurrence of neglected malignant bone tumors such as neglected osteosarcoma in developing countries as in our case is due to a combination of several epidemiological factors such as; unawareness, low socio-economic status and lack of diagnostic facilities and therapeutic options. The lack of awareness of the severity of the illness on the part of the patients combined with patients' cultural or religious beliefs and preferences also contribute to a large extend to the delay in presentation for medical care. The effect of the extremity tumors, especially in lower limbs significantly impaired the mobility and decreased the patient's quality of life.[5]

The most frequent sites of osteosarcoma include the metaphyseal bone of the distal femur, the proximal tibia, and the proximal humerus. The patients with neglected osteosarcoma typically present with enlargement of the tumor with either ulceration of the skin or pain. Initially, pain may be related to activity, but progresses to rest pain or even night pain which unfortunately is commonly attributed to "growing pains." The median time from onset of symptoms to diagnosis is 4 months, though significant variability exists especially in underdeveloped or developing counties. Mild blunt trauma is often reported as an antecedent event but currently, there is no significant correlation between trauma and osteosarcoma.[2]

Radiological imaging is the gold standard diagnostic options for assessing extend and stage of malignant bony tumours such as osteosarcoma. Plain radiographs of the involved bone and adjacent joint have become a popular option due to low cost with high diagnostic values. Typical osteosarcoma radiographs appear as a mixed lytic and radiodense lesion adjacent to the metaphyseal bone in an eccentric manner. Cortical destruction and periosteal reaction are common, and typically manifest in a sunburst pattern. There is frequently mass extension into surrounding tissue.[2] In our institution, Computer Tomography (CT) imaging is not routinely perform and thus tumour staging for distant metastases are based on clinical examination, an ultrasound exam of the abdomen, and plain radiographs of the chest. Fine needle aspiration cytology (FNAC) is a mandatory diagnostic tool in diagnosing bone tumor lesions. The advantages of FNAC for diagnosing bone tumor includes simple, inexpensive, does not require general anesthesia, allows preliminary diagnosis within 15–20 min of aspiration with minimal possibility of seeding the tumor cells. The sensitivity of FNAC for categorizing bone tumors into benign and malignant was 94.7%. For soft tissue tumors, FNAC had a sensitivity of 90.9% and specificity was 100%.[6] In majority cases of osteosarcoma, presence of osteoid or osteoid-like material together with pleomorphic cells is highly diagnostic of osteosarcoma. In the majority of osteosarcoma especially in the neglected case, the combined investigation of clinical and radiological data together with the FNAC result has been

sufficient for making treatment decisions.[7]

The current standard of care for neglected osteosarcoma is the combination of chemotherapy, complete surgical resection of the primary disease, evaluation of possible metastasis from the primary tumor and its related treatment .[8,9] Complete surgical resection of the tumor should be the goal for curative treatment in osteosarcoma regardless of the location. In the malignant bone tumor, amputation has been reported as a good oncologic procedure, especially in neglected case. Despite this, amputation is often misconstrued to be a procedure with poor functional results. With the use of excellent prosthesis patients with osteosarcoma who have undergone lower limb amputations showed the ability to sustain a much higher activity level, including impact activities such as running.[2,8,10]

In the developing countries, treatment options are relatively limited with far less access to the more advanced treatment care compared to a developed healthcare system. The challenge in treating neglected osteosarcoma case in developing countries not just influenced by the a combination of advanced pathology due to delayed treatment and unawareness of the patient, poor access to investigative modalities either financial or lack of diagnostic tools, cultural reluctance to comply with Western treatment related to local culture, limited access to adjuvant therapy and lack of modern surgical limb salvage techniques.[11]

CONCLUSION

The health-seeking behavior in neglected osteosarcoma patient resulted from the combination of unawareness, low education, low socio-economic level, unable to access healthcare provider and others. An adequate diagnostic and early treatment pathway is needed for patients presenting with neglected malignant bone tumours in order to improve the quality of life of these patients.

DISCLOSURE

None of the authors have direct or financial conflicts of interest with this paper and material contained herein. Authors also acknowledged that consent has been obtained to publish these images.

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