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Case Report

Proximal Tibial Osteosarcoma in Pediatric Patient: A Case Report

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ABSTRACT

Background: The most frequent primary malignant bone tumor in children is osteosarcoma. Patients with osteosarcoma are currently treated with a combination of neoadjuvant and adjuvant chemotherapy as well as surgery, either limb-sparing surgery or limb-ablation surgery. How to manage the remaining bone growth during the limb-sparing procedure in youngsters is one of the challenges. Unfortunately, amputation is still many perform for OS patients. The purpose of this study was to inform and educate doctors about the value of early OS diagnosis and treatment in pediatric patients.
Case Presentation: We presented case from a male 16 years old Balinese complaint of pain on her left knee 12 months ago. The pain was continuous and got worse at night. The patient went to a traditional masseuse and was given topical herbal ointment. Three months later, a lump appeared on her knee at the size of a small ball and getting bigger. We did a review slide of the previous biopsy sample by the pathology of anatomy in our hospital. The result came out as osteosarcoma with osteoblastic cells. We performed to amputated the limb-affected tumor following wound care routinely controlled.
Results: Intraoperatively, we performed wide excision, without patellar tendon, medial collateral ligament, and cruciate ligament preserve. We amputated the limb-affected tumor. The patient routinely controlled to our outpatient clinic every week. Unfortunately, the patient was died after 3 weeks of admission.
Conclusion: To reduce the risk of misdiagnosis or delayed diagnosis of osteosarcoma, every source of knee pain with a mass in pediatric age groups should be thoroughly explored with a high index of suspicion for malignant lesions. Therefore, amputation is a cutting-edge osteosarcoma treatment that can be avoided if the patient can be identified early and treated quickly in pediatric oncology malignant instances.

Keywords: osteosarcoma, long bone, diagnosis, tibia, pediatric

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BACKGROUND

The most frequent primary malignant bone tumor in children is osteosarcoma (OS) (Prabowo et al., 2020). Proximal tibia and the distal femur are the two most frequent sites (Mesregah, 2020). A primary malignant bone tumor that affects 3.4 million people annually worldwide is osteosarcoma. According to the significant bone growth that occurs during adolescence and

early adulthood, osteosarcoma is most frequently diagnosed during these years. The age group from 15 to 19 years old has the highest yearly incidence of 8.9 per million, with ages 10 to 14 years old having a somewhat lower rate. Only 1% of the nearly 3000 patients with osteosarcoma studied in a recent study were under the age of five (Abrams et al., 2013). In the 20 th century, the five-year survival rate for classic OS was 20%, but in 1970, the use of adjuvant chemotherapy in the treatment of OS increased survival rates to 50% (Blasius et al., 2022). In the middle of the 1970s, amputation was the surgical option for high-grade OS. Modern operating systems put more attention on chemotherapy and limb preservation around 1990, which led to a current survival rate of more than 65% (Sheng et al., 2021).

Patients with osteosarcoma are currently treated with a combination of neoadjuvant and adjuvant chemotherapy as well as surgery, either limb-sparing surgery or limb-ablation surgery (Blasius et al., 2022; Sheng et al., 2021). How to manage the remaining bone growth during the limb-sparing procedure in youngsters is one of the challenges (Lee, 2015). Unfortunately, amputation is still many perform for OS patients (Lee, 2015). Amputation has typically been the surgical method used to treat osteosarcoma. The great majority of patients will not be cured by even the most ablative of procedures, it was realized as early as 1879. The importance of chemotherapy in prolonging patients' lives with osteosarcoma was not recognized until the 1970s. While the addition of chemotherapy has improved survival rates, it has been accompanied by a trend away from surgery (Tiwari, 2012). The purpose of this study was to inform and educate doctors about the value of early OS diagnosis and treatment in pediatric patients.

CASE PRESENTATION

A male 16 years old balinese presented with pain on her left knee since 12 months ago. The pain was continuous and got worse at night. The patient went to a traditional masseuse and was given topical herbal pointment. Three months later, a lump appeared on her knee at a size of small ball and getting bigger. The patient then went to a general surgeon and a histopathological examination demonstrated bone tumor and the patient had previously done several chemotherapy sessions and discontinued the schedule in the past year. She was subsequently referred to our hospital. The patient was student at senior high school. From the medical record and family history that related to his diagnosis were absence. His nutrition intake was good, and defecation or urinary problem were within normal range.

Physical examination demonstrated a lump on the anterior part of the left proximal tibia with previous surgical biopsy scar and venectation. The lump had hard consistency with an ill-defined border, smooth surface, immobile, and pain (visual analogue scale 3-4). The size circumferential of the leg was 39 cm compared to the contralateral (see Figure 1). Neurovascular was good. The range of movement of both of the knee and ankle was normal. Plain radiography suggested mixed lytic and blastic bony lesion in metaphyseal area of the proximal tibia, with ill-defined margin, wide transitional zone, periosteal reaction, matrix osteoid and also soft tissue involvement (see Figure 2). Plain radiography of chest showed no sign of pulmonary metastasis. Contrast-enhanced magnetic resonance imaging (MRI) showed heterogeneous mass and no neurovascular bundle involvement. Laboratory examination demonstrated increased erythrocyte

sediment rate 53 mm/hour (N: < 15), and lactate dehydrogenase 880 U/L (N: < 480).

RESULTS

We did a review slide of the previous biopsy sample by the pathology of anatomy in our hospital. The result came out as osteosarcoma with osteoblastic cells. Then the patient underwent chemotherapy neo adjuvant followed with surgery by amputation procedure, wide excision. The patient underwent chemotherapy neoadjuvant for 4 cycles weeks with regiments Cisplatin and Doxorubicin.

Intraoperatively, we performed wide excision, without patellar tendon, medial collateral ligament, and cruciate ligament preserve. We amputated the limb-affected tumor (see Figure 3 & 4), The patient routinely controlled to our outpatient clinic every weeks. Unfortunately, the patient was died after 3 weeks admission.

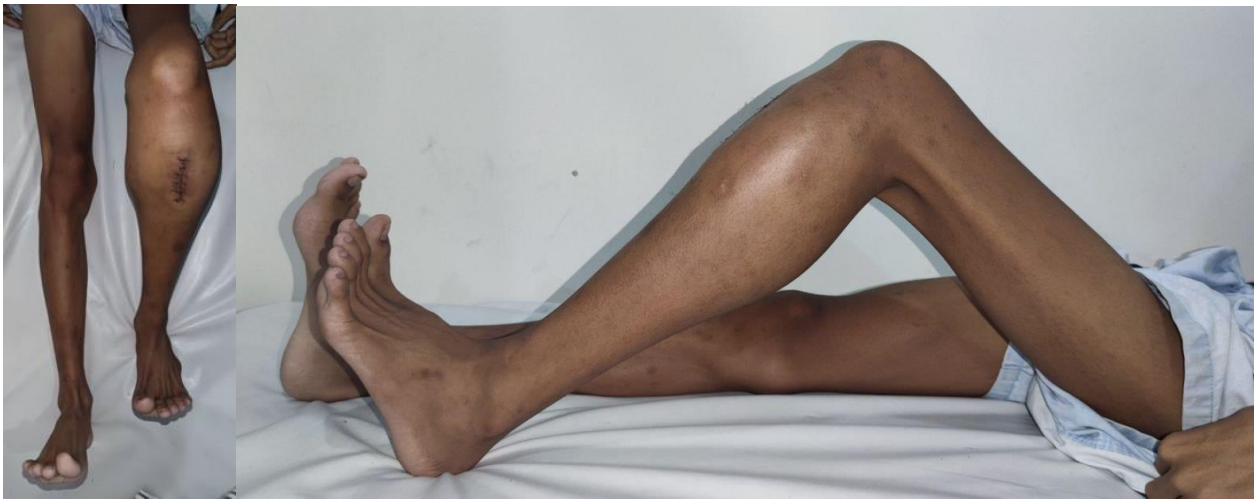

Figure 1. Clinical examination showed a mass on the left proximal tibia

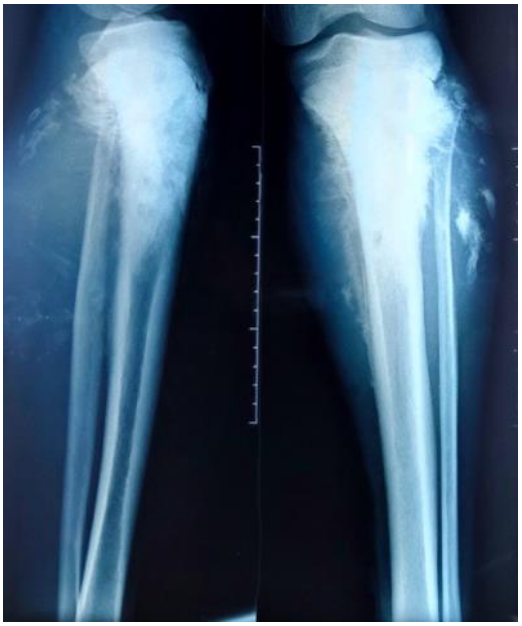

Figure 2. Radiograph examination (AP & lateral) showed on the left tibial

Figure 3. Post Amputation, the size was average 60 cm x 55 cm with ischaemic distally of the limb affected (12 months after first clinical presented)

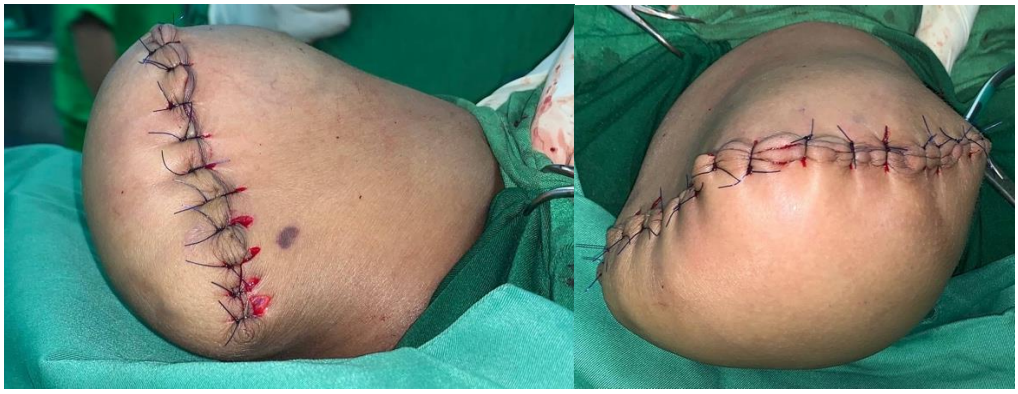

Figure 4. Post amputation affected limb

DISCUSSION

Osteosarcoma (OS) is a malignant tumor that originates in the mesenchymal tissue (which constitute spindle-shaped stromal cells that can produce bone-like tissues), and it accounts for 20% of all cases of primary malignant bone tumors in the world (Raymond and Jaffe, 2009). Both types of cancers develop as a result of genetic alterations, which are a crucial factor. Patients with hereditary retinoblastoma have up to 1000 times more risk of developing an OS due to germline mutations in the Rb gene (Sheng et al., 2021). Moreover, the loss of heterozygosity, structural rearrangements or specific mutations of the gene are present in 60- 70% of sporadic OS (Ritter and Bielack., 2010). Abnormalities in genes that regulate the cell cycle, such as p53, p16, Cyclin D1, MDM2, among others, have been implycated in the genesis of non-hereditary OS (Broadhead et al., 2011).

Osteosarcoma of the limb bones is mostly diagnosed through a combination of pathological, radiological, and clinical examination. The relatively constant cooperation of the multidisciplinary team participating in the diagnostic process can make the diagnosis and differential diagnosis more accurate (Reddy et al., 2015). Clinically, the onset of the disease is characterized mainly by local pain and swelling, and occasionally by joint dysfunction. A few patients have also been treated for pathological fractures. The symptoms of growth pain and trauma are confounding, but the degree of malignancy is high (Raymond and Jaffe, 2009). Notably, nearly 10–20% of the patients are affected by measurable metastatic disease before actual onset, the most common site being the lungs (85%), followed by the bones (8–10%) and, occasionally, the lymph nodes. The remaining 80–90% of the patients can be considered to possess subclinical or micrometastases, which cannot be detected accurately by using the presently available diagnostic methods (Anderson., 2016).

Theoretically, high-grade osteosarcoma cases can be distinguished from low-grade instances by a big mass with irregular borders, a destructive, mixed, lytic, and blastic character, and frequent soft tissue invasion. Low-grade cases are typically sclerotic and originate from the cortical aspect. Parosteal low-grade osteosarcomas, which occur at a rate of 3-5%, and intramedullary low-grade osteosarcomas, which occur at a rate of 1%, comprise up low-grade osteosarcomas (Narang et al., 2018). It is crucial to distinguish between high-grade and low-grade osteosarcomas because the latter has a better prognosis. It is also crucial to accurately assess the overlap between benign lesions in the differential diagnosis and to create a multidisciplinary treatment plan for the patient's diagnosis, treatment, and follow-up (Nath Srivastava et al., 2021).

The goal of treatment is to achieve local control of the tumor and systemic control of the disease. Chemotherapy has been the main factor in the increased survival rates seen over the last several decades for patients with osteosarcoma (Knollmann-Ritschel et al., 2017). High-grade osteosarcoma must be considered as a systemic condition. For instance, metastatic disease appeared in 80–90% of patients with seemingly isolated osteosarcoma treated with amputation alone within the first two years after surgery. This suggests that the majority of individuals have microscopic metastatic illness that is undetectable at the time of initial diagnosis. The rates of disease-free survival have dramatically increased as a result of the addition of adjuvant chemotherapy to the treatment of this micrometastatic disease. The chance of being disease-free following osteosarcoma amputation was less than 20% before the advent of chemotherapy; the present probability is estimated to be between 65 and 80% (Herring, 2002).

This case aims to demonstrate that rapid identification and treatment of OS are essential components of successful strategies. Although the surgical management of OS of the proximal tibia has received extensive research, the information on patient survival and function following limb salvage and amputation for OS in this region has been inconsistent. Mavrogenis et al., compared survival, local recurrence, function, and complications of patients with osteosarcoma of the distal or proximal tibia treated with limb salvage or amputation. Function was superior in patients with limb salvage compared to those who had amputations in terms of survival, local recurrence, and comorbidities (Mavrogenis et al., 2012).

Amputation is a preferable in most of early osteosarcoma, especially in cases where the other treatment modalities is found inadequate. (Marulanda et al., 2008). Xu et al stated that, the aim of surgical treatment for OS has evolved from saving lives to maximizing the functions of the affected limbs (Xu et al., 2020). Limb salvage surgery refers to the surgical procedure to restore bone and joint function after extensive resection of malignant bone tumors of the limbs (Hasley et al., 2018). Levin et al stated that, the key to the operation is to select the appropriate boundary (Levin et al., 2017). With the recent popularization of comprehensive limb salvage therapy in combination with neoadjuvant chemotherapy, limb salvage surgery has been used more often in clinical applications. In fact, limb salvage surgery is an option for 80–95 percent of patients with soft tissue sarcoma of the bones and limbs. Although local amputation recurrences and limb salvage have the same incidence rates, limb salvage patients had a greater 5-year survival rate (Levin et al., 2017). Limb salvage surgery preserves the patient's apparent integrity not only functionally, but also externally (Simpson and Brown, 2018). In OS surgery should completely remove the lesion to avoid local recurrence and distant metastasis. If the lesion is not completely removed during the operation, the local recurrence rate can be as high as 25% (Zhao et al., 2019).

OS is a cancer that spreads quickly, and the prognosis depends on whether it is discovered before it metastasizes (Ferguson and Turner, 2018). It has been demonstrated, in particular, that misdiagnosis and the ensuing delay in treatment are linked to worse survival; given that metastasis alters prognostic outcomes to such an extent, it is crucial to detect cancer early in order to increase survival rates among the patient cohort (Werner, 2016). There are several factors that make this a tough feat to accomplish: (1) the ambiguous osteosarcoma clinical presentation, (2) lack of understanding of the diseases by primary care, and (3) the lack of multidisciplinary action, which can lead to misdiagnosis (Wang et al., 2019). Therefore, it is important to understand the most efficient technique that will permit an accurate diagnosis.

In summary, to reduce the risk of misdiagnosis or delayed diagnosis of osteosarcoma, every source of knee pain with a mass in pediatric age groups should be thoroughly explored with a high index of suspicion for malignant lesions. Therefore, amputation is a cutting-edge osteosarcoma treatment that can be avoided if the patient can be identified early and treated quickly in pediatric oncology malignant instances. Additionally, amputation plus neoadjuvant chemotherapy has a beneficial overall effect, enhancing prognosis. Unfortunately, osteosarcoma still has a poor prognosis. Therefore, the multidisciplinary treatment constituted an appropriate alternative in patients with osteosarcoma of the proximal tibia.

AUTHOR CONTRIBUTION

Komang Septian Sandiwidayat is an expert, conceptual, editing, reviewing. Febyan searches for literature, editing, reviewing.

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CONFLICT OF INTEREST

There are no conflicts of interest

REFERENCE

Abrams AK, Avedian RS, Marina N (2013). Treating pediatric osteosarcoma: recent clinical trial evidence. Clin. Invest. 3(10): 967–978. Doi: 10.4155/cli.13.8-9.

Anderson ME (2016). Update on Survival in Osteosarcoma. Orthop Clin North Am. 47(1): 283–292. Doi: 10.1016/j.ocl.2015.08.022.

Bläsius F, Delbrück H, Hildebrand F, Hofmann UK (2022). Surgical Treatment of Bone Sarcoma. Cancers. 14(11): 2694. Doi: 10.3390/cancers-14112694.

Broadhead ML, Clark JCM, Myers DE, Dass CR, Choong PFM (2011). The Molecular Pathogenesis of Osteosarcoma: A Review. Sarcoma. 1–12. Doi: 10.1155/2011/959248.

Ferguson JL, Turner SP (2018). Bone Cancer: Diagnosis and Treatment Principles. Am fam physician. 98(4): 205-213. PMID: 30215968

Hasley I, Gao Y, Blevins AE, Miller BJ (2018). The Significance of a 'Close' Margin in Extremity Sarcoma: A Systematic Review. Iowa Orthop J. 38: 123-130. PMID: 30104934

Herring J ed (2002). Malignant Bone Tumors, in Tachdjian's Pediatric Orthopaedics. 3rd ed. Philadelphia: Sunders.

Knollmann RBEC, Regula DP, Borowitz MJ, Conran R, Prystowsky MB.(2017). Pathology Competencies for Medical Education and Educational Cases. Acad. Pathol. 4:1-36. Doi: 10.1177/2374289517715040.

Lee JA (2015). Osteosarcoma in Korean children and adolescents. Korean J Pediatr. 58(4):123-8. Doi: 10.3345/kjp.2015.58.4.123.

Levin AS, Arkader A, Morris CD (2017). Reconstruction Following Tumor Resections in Skeletally Immature Patients. J Am Acad Orthop Surg. 25(3): 204–213. Doi: 10.5435/JAAOS-D-15-00619.

Marulanda GA, Henderson ER, Johnson DA, Letson GD, Cheong D (2008). Orthopedic Surgery Options for the Treatment of Primary Osteosarcoma. Cancer Control 15(1): 13–20. Doi: 10.1177/107327480801500103.

Mavrogenis AF, Abati CN, Romagnoli C, Ruggieri P (2012). Similar survival but better function for patients after limb salvage versus amputation for distal tibia osteosarcoma. Clin Orthop Relat Res. 470(6): 1735-1748. Doi: 10.1007/s11999-011-2238-7.

Hasan BZ, Mesregah MK (2020). Limb Salvage in Pediatrics with Malignant Bone Tumors:Team Approach to Orthopedic Oncology. EJMO. 4(1): 26-34. Doi: 10.14744/ejmo.2020.52-322.

Narang NC, Diwaker P, Narang S, Venugopal VK (2018). Low-Grade Central Osteosarcoma: Report of Two Unusual Morphologic Variants. Indian J Surg Oncol. 9(1): 74–78. Doi: 10.1007/s13193-017-0713-7.

Srivastava RN, Sharma P, Raj S, Raj L, Dwivedi M, Kumar D, Ali W (2021). An Update on the Diagnosis, Treatment and Prognosis in Osteosarcoma. Acta Sci. Cancer Biol. 5(9):28-36.

Prabowo Y, Primaputra MRA, Kodrat E (2020). Reconstruction of osteosarcoma of the proximal tibia using bone on polyethylene hemiarthroplasty knee joint system: A case report. Int. J. Surg. Case Rep . 72:188–96. Doi: 10.1016/j.ijscr.2020.05.013.

Raymond AK, Jaffe N (2009). Osteosarcoma Multidisciplinary Approach to the Management from the Pathologist's Perspective. Cancer Treat Res.

152: 63–84. Doi: 10.1007/978-1-4419-0284-9_4.

Reddy KI, Wafa H, Gaston CL, Grimer RJ, Abudu AT, Jeys LM, Carter SR, et al. (2015). Does amputation offer any survival benefit over limb salvage in osteosarcoma patients with poor chemonecrosis and close margins?. Bone Joint J. 97(1): 115–120. Doi: 10.1302/0301-620X.97B1.33924.

Ritter J, Bielack SS (2010). Osteosarcoma. Ann Oncol. 21(7): vii320–vii325. Doi: 10.1093/annonc/mdq276.

Sheng G, Gao Y, Yang Y, Wu H (2021). Osteosarcoma and Metastasis. Front. Oncol. 11: 780264. Doi: 10.3389/-fonc.2021.780264.

Simpson E, Brown HL (2018). Understanding osteosarcomas. JAAPA. 31-(8): 15–19. Doi: 10.1097/01.JAA.000-0541477.24116.8d.

Tiwari A (2012). Current concepts in surgical treatment of osteosarcoma. J Clin Orthop Trauma. 3(1):4-9. Doi: 10.1016/j.jcot.2012.04.004.

Wang J, Liu S, Shi J, Li J, Wang S, Liu H, Zhao S, et al. (2019). The Role of miRNA in the Diagnosis, Prognosis, and Treatment of Osteosarcoma. Cancer Biother. Radiopharm. 34(10): 605–613. Doi: 10.1089/cbr.2019.293-9.

Werner M (2016). Avoiding pitfalls in the diagnostics of bone neoplasms from pathologists point of view: Importance of reference pathology. Radiologe. 56(6): 536-42. Doi: 10.1007/-s00117-016-0117-7.

Xu M, Wang Z, Yu X, Lin JH, Hu YC (2020). Guideline for Limb-Salvage Treatment of Osteosarcoma. Orthop Surg. 12(4): 1021–1029. Doi: 10.1111/-os.12702.

Zhao J, Xu M, Zheng K, Yu X (2019). Limb salvage surgery with joint preservation for malignant humeral bone tumors: operative procedures and clinical application. BMC Surg. 19(1): 57. Doi: 10.1186/s12893-019-0519-3.