

Proximal Tibia Osteosarcoma Presenting Late: A Case Report

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Abstract: Osteosarcoma is a malignant tumor that primarily affects the long bones but can also involve other bones in the body. The age at presentation ranges from 15 to 25 years of age and predominantly affects males. In this paper we present a 25 years old male patient presenting with a 2 years history of pain and swelling around left knee with aggressive growth of tumor leading to restricted mobility and pathological fracture in left tibia with severe emaciation. He presented late to our hospital, 6 months after noticing wound over the left knee. Diagnosis of tumor is important especially in early stage for improving prognosis.

Keywords: Osteosarcoma, Retinoblastoma, Pathological fracture

1. Introduction

Osteosarcoma is a primary malignant tumour of the skeleton characterised by the direct formation of immature bone or osteoid tissue by the tumour cells [1]. The classic osteosarcoma is a rare (0.2% of all malignant tumours) highly malignant tumour, with an estimated incidence of 3 cases/million population/year [2]. Osteosarcoma arises predominantly in the long bones and rarely in the soft tissues. The age at presentation ranges from 15 to 25 years of age [3]. Years ago, all patients with osteosarcoma were treated by amputation but the cure rate was under 10% and almost all patients died within a year from diagnosis. Today, for localised osteosarcoma at onset (80% of cases) treated in specialized bone tumour centres with pre- and postoperative chemotherapy associated with surgery, the percentage of patients cured varies between 60% and 75% [4, 5].

2. Case Report

Mr. Nagaraj, a 25 years old male patient residing at Busavana kote, Jagaluru, Davangere district, Karnataka. He was admitted with chief complaints of pain around left knee since 2 years and unable to walk around since 3 months.

Patient was apparently alright 2 years back then he developed pain around left knee, insidious in onset gradually progressing, dull aching type of pain, which was present throughout the day and worsened at night, aggravated with bearing weight and partially relieved with rest and analgesics. Patient noticed a small swelling just below left knee, solitary which increased in size over a period of 7 months. 6 months back he developed ulcer over the swelling initially small in size over the centre of swelling which gradually increased to involve the whole mass, associated with blood tinged purulent discharge. He was bedridden since 3 months, and had history of significant weight loss and generalized wasting developing over a period of one year. He was diagnosed as retinoblastoma and underwent

enucleation of left eye due to mass lesion and associated loss of sight. Similar surgery was done for his elder sister in both eyes.

He presented late to our hospital, 6 months after noticing wound over the left knee. At the time of presentation, a solitary fungating mass of size 20cm*15cm over left leg anterior aspect below the knee joint with necrotic tissue in centre and pearly white mixed with haemorrhagic tissue in periphery with surrounding skin hyperpigmented. Margins being irregular and everted edges. The mass bleeds on touch. Left knee joint movements were painful and restricted.

Hb was 2.3 g/dl at presentation and after 6 transfusions of Packed cell volume of RBC it came to 11.6g/dl. Serum alkaline phosphatase was 239.4 U/L. Serum calcium 6.9mg/dl, serum phosphorous 3.1 mg/dl

Plain x ray left knee showed osteoblastic lesion with an area of irregular destruction in the metaphysis of proximal tibia with periosteal reaction and calcified new bone formation in matrix of tumor going centrifugally giving rise to sun burst appearance, and pathological fracture of proximal tibia.

Chest x ray showed no evidence of metastasis, wound debridement was done and biopsy was taken once the patient was fit to undergo the procedure. Histo pathological findings of Biopsy report showed infiltrating neoplastic lesion arranged in sheets with areas of necrosis. Cells were pleomorphic varying from round to spindle shape with high N:C ratio, vesicular nucleus and prominent nucleoli. Osteoid and fibroblastic areas surrounded by tumor cells, focally dilated vessels lined by tumor cells. All features suggestive of osteosarcoma.

Patient was then referred to specialized bone tumor centre in Bangalore for further management.



Figure 1: Clinical and radiological pictures at the time of presentation



Figure 2: Clinical and radiological pictures after wound debridement and biopsy

3. Discussion

Osteosarcoma is a highly malignant tumor derived from multipotential mesenchymal tissue characterised by malignant cells that produce osteoid matrix. It is second most common primary malignant tumor of bone after multiple myeloma. Peak incidences is in second decade and predominantly affects males. In more than 90% of cases it occur in the metaphysis of long tubular bones, but all bones can be affected, In decreasing order of incidence, distal femur 32%, proximal tibia 15%, proximal humerus 8%. Jaws are affected in elderly.

In most cases the cause of osteosarcoma is unknown. It occurs primarily in areas where bone growth takes place it originates where mutation occurs in rapidly dividing bone cells. There is a strong association of osteosarcoma with hereditary retinoblastoma (mutation in Rb gene) and with Li Fraumeni syndrome (in whom sequence of p53 is rearranged). There is increased incidence of osteosarcoma in patients who had radiotherapy as osteoprogenitor cells are sensitive to radiation.

Clinically predominantly pain is the first symptom to appear, 25% patients experience night pain. A large mass measuring over 5 cm with skin over swelling stretched and shiny with dilated veins and local rise of temperature having variable consistency. Restriction of motion in adjacent joints and

pathological fractures are seen in 5-10 % of cases. Constitutional symptoms often appear within months after the appearance of local symptoms, they are weight loss, pallor and anorexia.

The clinical course of untreated osteosarcoma is rapidly enlarging mass followed by pulmonary metastasis within 12-18 months and death within 18-24 months after the onset of local symptoms. Average delay from onset of symptoms to correct diagnosis is approximately 15 weeks in one study which includes average patient delay of 6 weeks (the time between onset of symptoms and initial physician encounter).

The tumors generally have violated the cortex of bone at the time of diagnosis. During active growth period while the epiphyseal cartilage is still intact it acts as a barrier to extension of tumor into epiphysis. After epiphyseal closure the tumor may extend into epiphysis but articular cartilage bars extension into joint. The tumor is variable in consistency and may be distinctly sclerotic; some soft areas are also present. Necrotic cyst formation and hemorrhagic areas are most commonly seen in soft portions of the tumor. The radiological features include osteoblastic lesions with soft tissue extension. As the periosteum is elevated a triangle of subperiosteal new bone formation occurs called as codman's triangle. Spicules of neoplastic bones are seen at right angle to long axis of bone along with blood vessels elevated by periosteum giving a sun-burst appearance.

Serum alkaline phosphatase is the biological marker elevated in 50% of cases of osteosarcoma helpful in determining the prognosis. An open biopsy is performed to confirm the diagnosis.

Treatment involves using chemotherapy to shrink the tumor size using cisplatin, adriamycin, methotrexate followed by En block curative resection of tumor (sacrificing common peroneal nerve and anterior peroneal artery) or amputation.

4. Conclusion

Osteosarcoma involves young population, and is a highly malignant and rapidly growing tumor which requires early diagnosis and treatment. Despite treatment consisting of wide or radical amputation approximately 80% of patients die as a result of distant metastasis usually within 2 years. With today's multiple agent chemotherapy regimen and appropriate surgical treatment most series report long term survival 60-75% of patients with high grade osteosarcoma without metastasis at initial presentation. So the patient was referred to specialized bone tumor centre once the diagnosis was confirmed by biopsy.

References

- [1] APA Picci, P. (2007). Osteosarcoma (Osteogenic sarcoma). Orphanet Journal of Rare Diseases, 2, 6. <http://doi.org/10.1186/1750-1172-2-6>
- [2] Durfee, R. A., Mohammed, M., & Luu, H. H. (2016). Review of Osteosarcoma and Current Management. Rheumatology and Therapy, 3(2), 221–243. <http://doi.org/10.1007/s40744-016-0046-y>
- [3] Mirabello, L., Troisi, R. J., & Savage, S. A. (2009). International osteosarcoma incidence patterns in children and adolescents, middle ages, and elderly persons. International Journal of Cancer. Journal International Du Cancer, 125(1), 229–234. <http://doi.org/10.1002/ijc.24320>
- [4] Isakoff MS, Bielack SS, Meltzer P, Gorlick R. Osteosarcoma: current treatment and a collaborative pathway to success. J Clin Oncol. 2015;33(27):3029–3035. [PMC free article] [PubMed]
- [5] Rosen G, Caparros B, Huvos AG, et al. Preoperative chemotherapy for osteogenic sarcoma: selection of postoperative adjuvant chemotherapy based on the response of the primary tumor to preoperative chemotherapy. Cancer. 1982;49(6):1221–1230. [PubMed]

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