

## Osteosarcoma in A Young Child- A Rare Cytological Case Report

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**Abstract :** The authors describe a case of osteosarcoma in a 9 years old male who presented with complaints of pain and swelling in Left leg in the OPD facility of RIMS, Ranchi, Jharkhand. Osteosarcoma is the most common malignancy in childhood and adolescence. However, it is very rare in children under 5 years of age. Although studies in young children are limited in number, they all underline high rate of amputation in this population, with conflicting results being recently reported regarding their progress.

**Keywords :** Osteosarcoma, Young children.

### I. Introduction

Osteosarcoma is defined as the primary malignant mesenchymal bone tumor where the malignant tumor cells directly form the osteoid or bone or both.<sup>1,2,3,4,5,6,7,8,9,10,11,12</sup> Demonstration of osteoid directly formed by the malignant cells in histopathology is essential for making the diagnosis of osteosarcoma.<sup>2,3</sup>

Although the exact cause of osteosarcoma is still unknown, defects in RB and p53 genes play an important role in the process. Patients with germline mutations in RB have approximately 1000-fold increased risk of osteosarcoma and similarly patients with Li-Fraumeni syndrome (germline p53 mutation) also have greatly elevated incidence of this tumor. Abnormalities in INK4a, which encodes p16 (a cell cycle regulator) and p14 (which aids and abets p53 function) are also seen. It is noteworthy that osteosarcoma occurs more commonly at sites of bone growth, presumably because proliferation makes osteoblastic cells to acquire mutations that could lead to transformation.<sup>1</sup> Radiation too has been implicated in causation.<sup>1,2</sup> The risk of developing postradiation osteosarcoma correlates with radiation dose and use of electrophilic chemotherapeutic agents.<sup>13,14,15</sup> An etiological relationship has not been proven in prosthesis and metal hardware associated osteosarcomas.<sup>16</sup>

Bone tumors make up about 3–5% of childhood cancers and less than 1% of cancers in adults.<sup>1–4</sup> Of these, osteosarcoma is the most commonly diagnosed primary malignant bone tumor.<sup>1,5–7</sup> Individuals with localized osteosarcoma have an average 5-year survival of about 80% but those with metastatic disease have much worse outcomes.<sup>8</sup> Its incidence is bimodally distributed by age with peaks in adolescence and in the elderly.<sup>1,8–12</sup> Osteosarcoma incidence in childhood and adolescence appears to be relatively consistent throughout the world,<sup>5,13–16</sup> however, international comparisons of incidence for other age groups have not been described. We report a case of a 9 year old Indian boy who presented with bone swelling and pain in right leg and was diagnosed with osteosarcoma on radiological and pathological examinations.

### II. Case report

A 9 year old male presented to OPD facility of RIMS, Ranchi with complains of pain and swelling in right upper leg. There was no history of trauma or prior radiation therapy. X-ray anteroposterior and lateral views of proximal tibia and knee joint showing metadiaphyseal osteosarcoma of tibia with sclerosis, cortical destruction on posteromedial side and new bone formation in soft tissue. Fine needle aspiration cytology was done with proper aseptic precautions which revealed pleomorphic, multinucleated, round tumor cells resembling osteoblasts along with atypical mitotic figures. Clumps of amorphous, faintly eosinophilic material in background were seen on H & E staining.

### III. Discussion

Osteosarcoma is the most common primary osseous malignancy excluding malignant neoplasms of marrow origin (myeloma, lymphoma and leukemia) and accounts for approximately 20% of bone cancers. It predominantly affects patients younger than 20 years and mainly occurs in the long bones of the extremities, the most common being the metaphyseal area around the knee. These are classified as primary (central or surface)

and secondary osteosarcomas arising in preexisting conditions. The conventional plain radiograph is the best for probable diagnosis as it describes features like sun burst appearance, Codman's triangle, new bone formation in soft tissues along with permeative pattern of destruction of the bone and other characteristics for specific subtypes of osteosarcomas. X-ray chest can detect metastasis in the lungs, but computerized tomography (CT) scan of the thorax is more helpful. Magnetic resonance imaging (MRI) of the lesion delineates its extent into the soft tissues, the medullary canal, the joint, skip lesions and the proximity of the tumor to the neurovascular structures. Tc99 bone scan detects the osseous metastases. Positron Emission Tomography (PET) is used for metastatic workup and/or local recurrence after resection. The role of biochemical markers like alkaline phosphatase and lactate dehydrogenase is pertinent for prognosis and treatment response. The biopsy confirms the diagnosis and reveals the grade of the tumor. Enneking system for staging malignant musculoskeletal tumors and American Joint Committee on Cancer (AJCC) staging systems are most commonly used for extremity sarcomas.

The plain radiograph provides the best clue to the diagnosis and MRI the local extent. Thorax CT scan and Tc99 bone scan are used for the detection of lung and bony metastasis respectively. Biopsy should be performed after complete history, clinical examination and imaging. It confirms the diagnosis, reveals specific type and furnishes the grade of the tumor. It is performed by either an open (incisional) or a closed method. Closed biopsy is performed as fine needle aspiration cytology (FNAC) and core needle biopsy.<sup>19</sup> Open or incisional biopsy is performed through a small incision and has the major advantage of obtaining adequate amount of sample for histopathology as well as for ancillary studies like immunohistochemistry (IHC) and genetic studies. But it takes more time and requires operation theatre set-up with instruments. There are more chances of contamination of normal soft tissue by tumor cells through an impending hematoma and also other complications like infection and wound problems posing greater morbidity. Further, there is more cost to the patient as it may require short stay in the hospital. However, if performed meticulously and properly, the complications can be reduced markedly almost comparable to those of a core needle biopsy.<sup>19</sup> Percutaneous core needle biopsy has now evolved as a better, safe and accurate method for diagnosing of bone tumors. It is performed through a small stab using the Jamshidi needle and taking multiple cores from the representative part of the tumor . It is less extensive and less time consuming outpatient procedure performed safely and quickly under local anesthesia and is cost effective. There is minimal soft tissue trauma with less contamination of normal tissue by the tumor cells around the tract of the needle which is easily excisable during the limb salvage surgery. It is very suitable for deep and difficult areas like the pelvis and spine<sup>17,18</sup>. The efficacy and accuracy can be further increased by performing this under image guidance i.e. under CT scan, MRI or ultrasonography. The recent literature advocates core needle biopsy as it provides adequate amount of sample for the diagnosis and the ancillary studies, and has less number of complications<sup>20,21,22,23,24,25,26,27,28,29,30,31,32</sup>. After clinical, radiological and the histopathological examinations the tumor can be staged adequately . It is pertinent to mention that the patient should be immediately referred to the treating specialist centre for early diagnosis and treatment as this can make limb salvage possible in large number of patients. The urgent need of the MDT (multidisciplinary team) for the better outcome in all musculoskeletal sarcomas can not be over-emphasised. However, the biological behavior of osteosarcoma is yet to be fully understood.

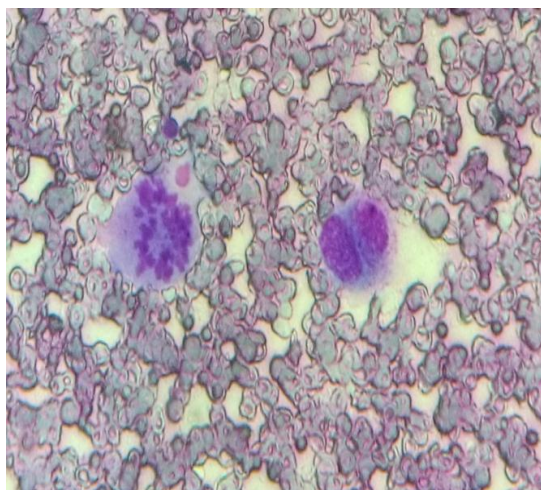
#### IV. Figures



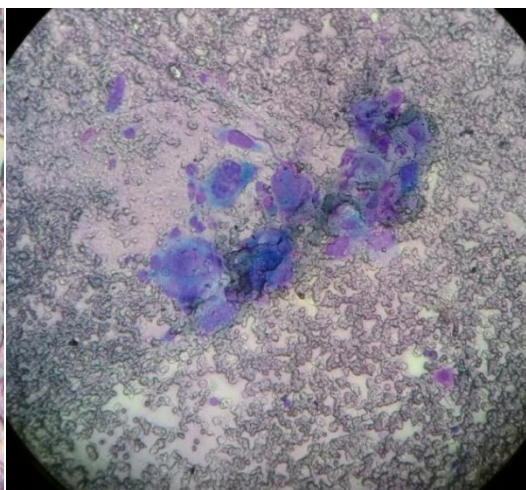
**Fig 1:-** Shows cortical bone destruction of tibia.



**Fig 2:-** Shows sclerosis and bone formation in soft tissue



**Fig 3:-** Malignant cells with macronucleoli, the fragments of osteoblast.



**Fig 4:-** Atypical mitotic figure with binucleate cells of pink amorphous material represent osteoid.

## V. CONCLUSION

Osteosarcoma is the most common type of bone cancer in children, typically occurring after the age of 10 years. It is extremely rare in children before the age of 5 years. Genetics can play a role in whether a patient develops osteosarcoma. Work up includes blood tests, radiological examinations including plain X-rays, CT chest and whole body imaging. A biopsy is always required to make the diagnosis. Core needle biopsy is preferred, but some times an open biopsy is required to obtain sufficient tissue to make diagnosis.

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