Diaphyseal Small Cell Osteosarcoma: A Case Report And Review Of Literature

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Diaphyseal Small Cell Osteosarcoma: A Case Report And Review Of Literature

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Introduction

Osteosarcoma is commonest primary tumor of the bone. Within its strict histologic definition, osteosarcoma consists of a group of lesions with considerable diversity in histologic features and grade. It may occur anywhere around the bone starting from intramedullary or intracortical to periosteal and extraosseous. It mostly occurs in the long bones in the second decade of life. It most commonly presents with pain and soft tissue swelling of 1-8 months duration as intramedullary type. Pathologic fractures are present in only 5% of cases. With combination chemotherapy and surgery, cure rates as high as 70-80% have been obtained when prior to this, only 10-20% 5 year survival rates were achieved. Conventional intramedullary osteosarcomas comprise 90% of all osteosarcomas but there are several other variants which lead to difficulty in diagnosis. Small cell osteosarcoma is the rare variety of it. Here, we are presenting a rare case of diaphyseal small cell osteosarcoma in a young male patient.

Case Report(s)

A 21 year old young male presented in our outpatient department with the chief complaints of pain and fever followed by swelling in the left thigh. The duration of the complaints was one month. The swelling was tender and mainly located over the medial aspect of the thigh. It was around 9cm * 4cm in size. Skin over the swelling was freely mobile. It was firm in consistency and no crepitation was present. Inguinal lymph nodes were not palpable. Distal neurovascular functions were normal. Detailed workup was done thereafter, including x rays. The x ray pictures showed a diaphyseal eccentric located tumor with sclerosis. The pattern of the destruction was not typical and sun-burst appearance and codman reaction were present. Clinical diagnosis of ewing sarcoma was made owing to its location. The patient was having chest metastasis in CECT chest. FNAC was done which showed multiple pleomorphic small cells with osteoid formation. The histopathological diagnosis was small cell osteosarcoma. Patient refused neoadjuvant chemotherapy. After staging of the tumor which was enneking stage 3, plan of high above knee amputation was made. Surgery was performed and postoperative rehabilitation of the patient was done. Patient does not having any complaints after 8 month of the follow up. Patient is walking with the prosthesis.

Discussion

Small cell tumors have several different families. Small cell variety in osteosarcoma has been confirmed by various studies. It is also known as multipotent sarcoma of the bone. All the features are same as intramedullary metaphyseal osteosarcoma except its location. It is diaphyseal in location. Formation of the new bone is one of the most important clues in the radiological pictures. This is important feature to differentiate it from ewing sarcoma. Another feature for detection is formation of osteoid in the biopsy material. Histopathology of both the tumours is more or less same except reactive bone sclerosis and soft tissue mineralization can be seen in Ewing's sarcoma, but the form of the soft tissue mineralization is that of laminated periosteal new bone. This is in contrast to the mineralized tumor matrix seen in the soft tissues or intramedullary compartment with osteosarcoma. The small cell osteosarcoma composed of small round cells which are similar to ewing sarcoma. But these small cells are more pleomorphic than in ewing sarcoma. These cells are also found to have spindle nuclei. There are three subtypes of small cell osteosarcoma which has been described by Ayala et al.

The term "small cell" is quite arbitrary. Most clinician will have an image of small cell tumors but the spectrum is very large. A diagnosis of small cell osteosarcoma is considered only if osteoid production was seen within the neoplasm. Sometime the amount of the osteoid produced scanty. FNAC is the most common tool to differentiate it from ewing sarcoma. Spindling of the tumor cells rule out the possibility of ewing sarcoma. Sometimes, other small cell malignancy like lymphoma and metastatic tumors can also cause problem in the diagnosis but the presentation is usually different.

The treatment and prognosis of osteogenic sarcoma depend very largely on the extent of the tumor. Localized, non-metastatic tumors in relatively
accessible sites such as the knee, hip, and shoulder generally has a very good to excellent prognosis. Very large tumors and tumors in difficult sites such as the pelvis and spine have a less favorable prognosis due to the difficulty with completely removing the tumor by surgical techniques. Once clinically evident metastasis occurs, the prognosis is substantially diminished. Tumors that present with metastasis already present or tumors where metastasis is discovered after the initial treatment have a poor prognosis. Surgical removal of metastasis coupled with chemotherapy may lead to improved survival.

References

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Illustrations

Illustration 1

X ray showing small cell osteosarcoma
Showing osteoid matrix and codman reaction

![X ray image showing small cell osteosarcoma](image-url)
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