

Juxtacortical Osteosarcoma-Imaging Spectrum

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Abstract

Juxtacortical Osteosarcoma (OS) presents with three sub-types. These include parosteal OS, periosteal OS, and High-grade surface OS. Imaging diagnosis of sub-types of Juxtacortical osteosarcoma is difficult. However, an analysis of 14 cases of these sub-types presents a spectrum of findings in each entity. The analysis of the Site, Transitional zone, Age, Matrix mineralization, Periosteal reaction, and Soft tissue component (STAMPS) are essential in making the final diagnosis and differential diagnosis.

Keywords: Juxtacortical Osteosarcoma, Parosteal, Periosteal, High grade surface sarcoma, Imaging findings.

Introduction

Osteosarcoma (OS) accounts for 15% of all primary bone tumors. Primary OS can be of various sub-types: high grade, low grade, small cell osteosarcoma, telangiectatic, gnathic, osteosarcomatosis, extra skeletal, surface (Juxta-cortical) osteosarcoma and secondary osteosarcoma. Juxtacortical / surface osteosarcoma accounts for 4-10 % of all osteosarcomas [1]. Based on the nature of tumor, clinical, radiological, and pathological findings, juxta-articular OS can be classified into parosteal, periosteal and high-grade surface osteosarcoma. In larger studies the age of diagnosis ranges from 8 to 70 years with a mean age of 25 years and with about 66% being males [2,3]. Surface OS was first described by Francis et al in 1964; but it was in 1984, Wold et al reported the first series of this rare tumor [2,4]. Majority of osteosarcomas arising from surface are low or intermediate grade and have relatively good prognosis as compared to conventional OS after wide surgical excision [5]. Pathologically parosteal OS arises from outer layer of periosteum and its cells are predominantly fibroblastic and these tumors are of low grade. Periosteal Sarcoma arises from germinative layer and cells are chondroblastic and are of intermediate grade tumors. High grade surface OS arises from the surface and have cells of high mitotic activity similar to conventional OS. Parosteal OS is treated by surgical excision without neoadjuvant chemotherapy whereas the High-grade surface OS has poor prognosis similar to conventional OS and needs neoadjuvant chemotherapy as standard care, since periosteal OS is of intermediate grade the role of chemotherapy is controversial.

The characteristic imaging features of Juxtacortical OS subtypes in extremities are unique. The application of the mnemonic "STAMPS" namely, **S** for site, **T** for transitional zone, **A** for age and aggressive nature, **M** for matrix mineralization, **P** for periosteal reaction and **S** for soft tissue changes, helps in analysing the imaging findings of these three entities.

The common site for parosteal osteosarcoma is the metaphysis of lower end of femur, upper end of tibia and upper end of humerus. The common site for periosteal Osteosarcoma is diaphysis/metaphysis of the long bones particularly tibia and femur. The common site for surface OS is Diaphysis/metaphysis of long bones. Femur is most common bone involved. Tibia is more commonly involved by Periosteal OS than Parosteal OS.

Transitional zone is wide in all the three types. Matrix mineralization is osteoid in all the three types and is excessive in parosteal osteosarcoma. Periosteal reaction of various types is seen in all the three entities. Soft tissue swelling is well appreciated on MRI and is larger in parosteal osteosarcoma.

Our Analysis of Juxtacortical Osteosarcomas

14 cases of histo-pathological examination (HPE) proven juxtacortical osteosarcoma are analysed. Male: Female ratio is 8:6. Majority were in age group of 20 years. There were 10 parosteal, 3 periosteal and 1 high-grade surface OS. The bones involved included femur, tibia, humerus and fibula in that order. 12 out of 14 were around knee joint. All the lesions were >1 cm in size. 10 out of 14 were surrounding the bone in less than 180 degrees and the rest 4 were more than 180 degrees. These findings agree with the published literature.

Parosteal Osteosarcomas

Parosteal osteosarcoma accounts for 5% of OS, occurring in 2-4th decade of life [6]. According to Okada et al the diagnostic criteria of Parosteal OS were:

1. Should have arisen from surface of bone
2. Histologically it should be grade 1 or 2 well differentiated tumor
3. Well-formed osteoid within a spindle cell stroma and medullary involvement if any should be <25% of medullary cavity (7).

Female predominates over male and long bones are common sites of involvement. However, axial skeleton may be involved. Posterior aspect of femur is most favoured location arising from fibrous layer of periosteum in 62% cases [7]. Metaphysis is a common site but diaphysis involvement is seen in 10% cases. Prognosis of Parosteal OS is better than conventional OS, being 86-91% as compared with latter 53-61% [8]. Radiologically the mass is lobulated, exophytic with central dense calcification (Figure 1, 2). A cleavage plane is noted between the mass and cortex except at the area of attachment. This is called string sign (Figure 1, 2) which may represent the uncalcified, uninvolved thickened periosteum and is seen in 30% of cases.

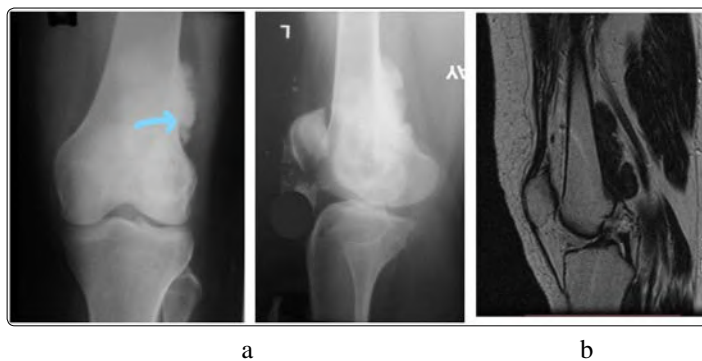


Figure 1ab: 22F, Parosteal Osteosarcoma of femur, a-Conventional, b-MRI

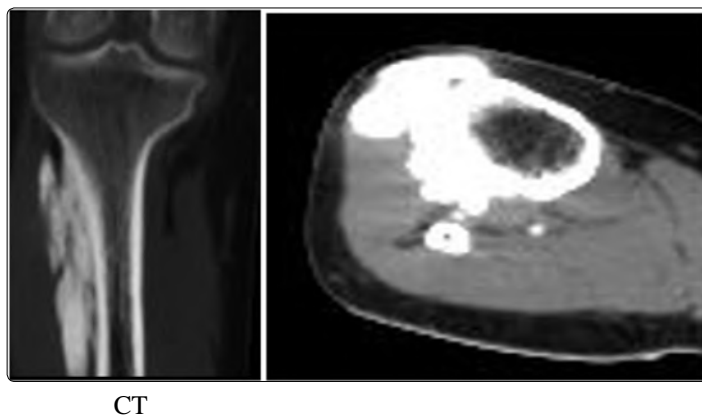


Figure 2: Parosteal Osteosarcoma of tibia, CT - String sign in coronal reformatted CT image.

However, this cleft may be obliterated with advancing tumor growth [9,10]. This lucent zone is better appreciated in CT Scan as compared with radiography (Figure 3).

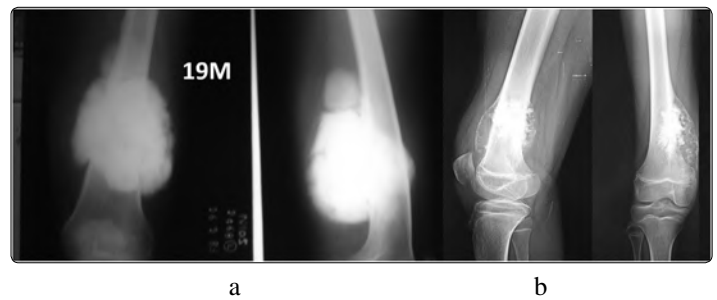


Figure 3ab: 19 M, Parosteal Osteosarcoma of femur, a- absence of string sign in advanced lesion, b-another case with CT

Okada et al found most of the tumor had a cortical attachment more than 1 cm and the mass tends to wrap around the involved bone as it grows (Figure 2,4 and 5) [7]. 80% of parosteal OS will involve 50% of circumference of native bone or less (Figure 4). Cortex may be normal, (Figure 1, 2) or thickened and rarely destroyed (Figure 3). Periosteal reaction is usually not seen. By definition surface OS have no intramedullary involvement, however intramedullary involvement and PR are seen in 10% cases (Figure 5). CT scan clearly defines the extent of tumor and cortical integrity (Figure 3,4).

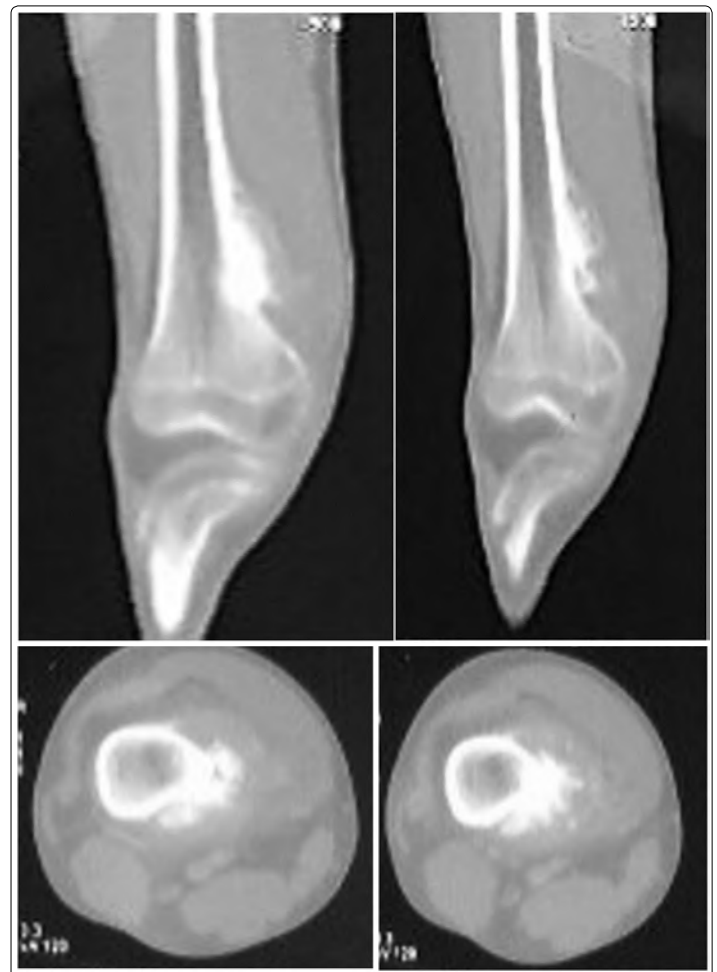


Figure 4: 12 M, Parosteal OS of tibia, tumor attachment is more than 1 cm and less than 180 degrees

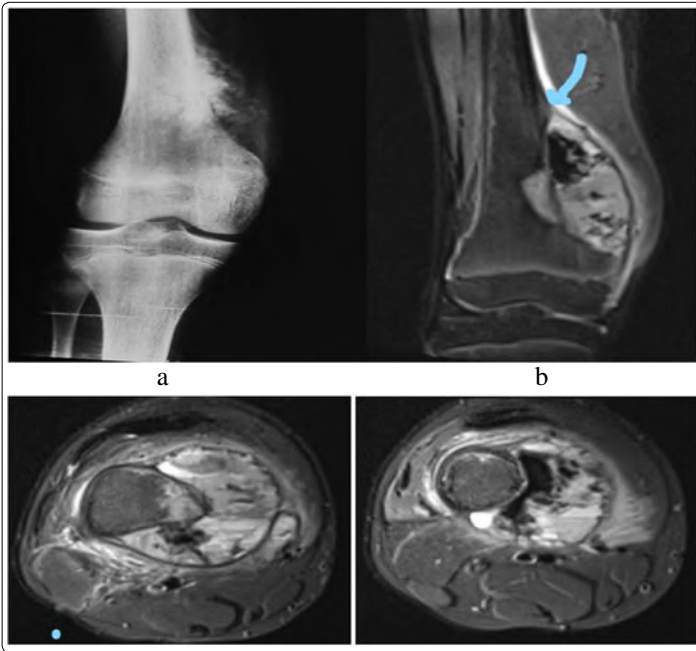


Figure 5abc: 13 M, a-conventional, b-MRI, Parosteal Osteosarcoma of femur, periosteal reaction, STIR hyper intense component and medullary involvement

The satellite lesion also could be detected on CT. MRI is useful to determine the size, site (Figure 6, 7) presence of osteoid, cartilaginous component, haemorrhage, necrosis and areas of high grade tumor (Figure 5). Hence MRI is useful to determine the site of biopsy. Intramedullary involvement is also appreciated by MRI (Figure 8). The tumor is hypo intense on both T1W, T2W images (Figure 1). This represents mineralised soft tissue component. A cartilaginous component appearing hyper intense on T2W images is observed in more than 50% of all parosteal osteosarcoma and 25% of it lies at periphery of tumor (Figure 5,7). When there is an unmineralized area of more than 1cm square or the lesion is predominantly high in T2W images, the tumor is more likely to be high grade [11].



Fig. 6: 20 M, Parosteal Osteosarcoma of the femur, no intramedullary involvement



Figure 7: 36 F, Parosteal Osteosarcoma of tibia, tumor more than 180 degrees surrounding the shaft

Pathologist and surgeon must recognise this cartilaginous component and should not confuse with osteochondroma. There may be dedifferentiation which is noted in 16-43% [7] and it may be Fibro sarcoma or malignant fibrous histiocytoma. The area of dedifferentiation is indicated by increased lysis and large soft tissue without calcification (Figure 9). Area of T2 hyper intensity within soft tissue of 3 square cm indicate high grade tumor (Figure 10). The presence of an ill-defined soft tissue mass within or adjacent to the ossified tumor suggests the area of dedifferentiation [9]. Lucent area within the osteoid mass in plain radiograph and CT Scan indicates dedifferentiated area of Parosteal osteosarcoma (Figure 9). PET CT shows high metabolic activity in dedifferentiated component as compared to rest which can be targeted for biopsy.

The close differential diagnosis is myositis ossificans. Here, the dense ossification seen at periphery in contrast to osteosarcoma where it is central. History of trauma is important in myositis ossificans. Cortical continuity with parent bone and area of

peripheral erosion are diagnostic clues to parosteal OS (Figure 8). Other differentials are osteochondroma and Periosteal chondroma. In osteochondroma the cortical and medullary continuity can be seen between the mass and parent bone. Rarely parosteal OS shows the continuity with adjacent cortex and medulla mimicking osteochondroma [12]. Periosteal chondroma causes saucerization as it arises from deep layer of periosteum. The mass typically arises from the surface of bone, exhibiting scalloping of the cortex and a well-defined margin between the tumor and bone.



Figure 8 ab: 54 F, Parosteal Osteosarcoma of femur, a-conventional, b-MRI, medullary involvement



Figure 10ab: 28 F, Parosteal Osteosarcoma of femur, a – conventional + MRI, b – Lungs with metastasis



Figure 9ab: 28 M, Parosteal Osteosarcoma of tibia, lucent areas (blue dots) within the tumor indicates dedifferentiation, a-conventional, b - CT

Periosteal Osteosarcoma

Periosteal OS accounts for 5% of osteosarcoma occurring in 2-4th decade of life [13]. Among the surface osteosarcomas, the periosteal OS occurs in a younger age group and are more common in males. They arise from deep germinative layer of periosteum and the tumor is predominantly cartilaginous. Diaphysis of long bones is a common site with fusiform involvement of the cortex (Figure 11). Cortex may be eroded and thickened. Periosteal reaction, large soft tissue mass and chondroid matrix are the common features of periosteal OS (Figure 11). Broad based soft tissue mass is noted which is inhomogeneous in attenuation/signal intensity (Figure 12). The tumor is hypo intense on T1 and T2W images depicting the osseous matrix. The chondroid matrix shows T2 hyperintensity and T1 low signal (Figure 13). There are ring and arc like enhancements within the tumor on contrast administration. The PR is perpendicular to the axis of bone giving sunburst pattern (Figure 12,13). Though intra-medullary extent is rare but several reports in literature have described it as a well-recognised feature [14].



Figure 11: 19 M, Periosteal Osteosarcoma of humerus, deep layer of periosteum is involved, Sunburst periosteal reaction, broad based soft tissue component

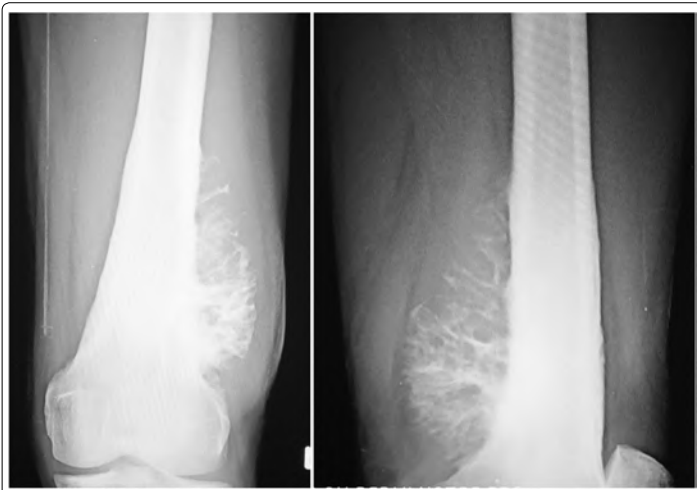


Figure 12: 34 M, Periosteal Osteosarcoma of femur, large broad-based soft tissue component

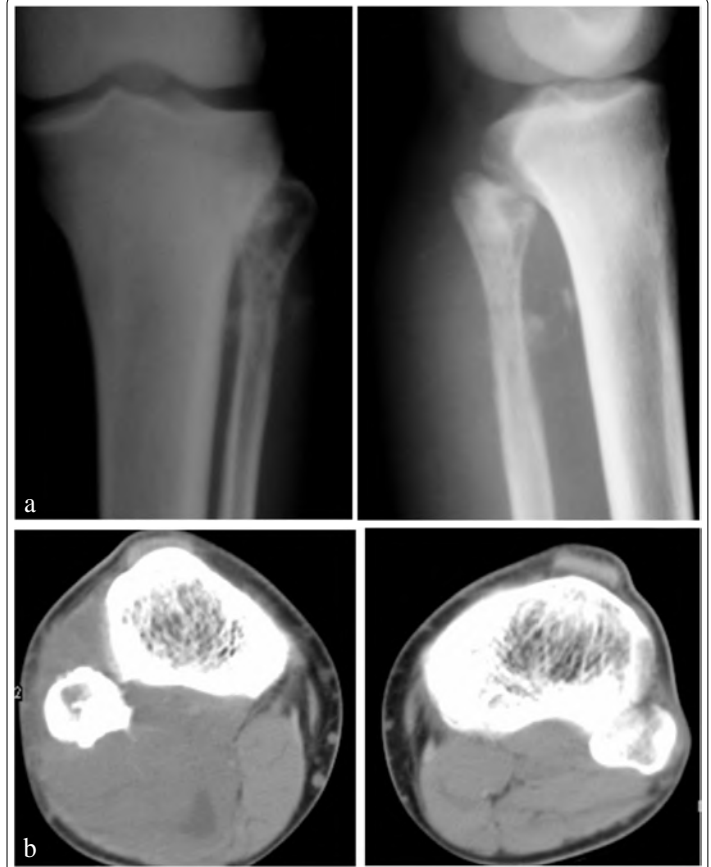


Figure 13ab: 20 M, Periosteal Osteosarcoma of fibula, large soft tissue mass, a-Conventional, lesion extending to the articular margin, b-CT showing large hypodense soft tissue mass along with PR.

Prognosis is better than conventional OS but poorer than parosteal OS and grade 2 tumor. The close differential is periosteal chondroid tumor which occurs in slightly elder age group. High grade surface osteosarcoma is another differential diagnosis. The latter involves the entire circumference of shaft and intramedullary involvement is common, which is not seen in periosteal osteosarcoma.

High Grade Surface Osteosarcoma

High grade surface OS is rare and accounts for 0.4% of all OS occurring in 2nd-3rd decade. This is the rarest of all juxtacortical OS and accounts for <1% of all OS [15]. It is high grade similar to that of conventional osteosarcoma. Diaphysis/metaphysis of long bones are common sites. Femur is the most common bone involved. The tumor is generally large, ranging from 4.5-22cms [15, 16]. Dense periosteal reaction in the form of cloudy new bone, cortical thickening, cortical destruction, and intramedullary invasion are seen in 8-48%. It involves the entire circumference of shaft and intramedullary extent is common (Figure 14). Murphey MD et al reported circumferential involvement of host bone in 28% and half of them were more than 50% [3]. The amount of matrix mineralisation and distribution varies from dense to moderate with fluffy, immature appearance predominantly at base of lesion [3]. Tumors with less ossification tend to be less differentiated than the tumors with more ossification (Figure 14).



a b

Figure 14ab: High grade Osteosarcoma of femur, tumor surrounding the shaft with periosteal reaction, a – Conventional, b - CT

Periosteal reaction is rare with no Codman's triangle and *sunray* speculation. Intramedullary extent is depicted by lower attenuation in marrow on CT and low signal intensity on T1, high signal on T2W MRI. Imaging wise sometimes it is difficult to differentiate from high grade conventional osteosarcoma. Epicentre of mass differentiates the two. This entity is also to be differentiated from Parosteal/Periosteal OS and Ewing's Sarcoma.

In a recent study of 18 cases of JOS by Nouri male: female ratio was 39:61 with a mean age of 25 years. Femur was involved in 11 (61%) and tibia in the rest. Parosteal, periosteal and high-grade surface OS were seen in 11,3 and 4 cases respectively [17]. These observations were similar to our present series of 14 cases.

Conclusion

Imaging analysis of sub-types of osteosarcomas of bone is difficult. However, some characteristic features differentiate the sub-types one from the other. Juxtacortical OS is divided into parosteal, periosteal and high-grade surface osteosarcoma. In the 14 cases presented, each one of them has distinct clinic pathological radiological features. Most of our findings coincide with the reported literature except the patients were younger in those with parosteal OS. Their recognition is important as prognosis and treatment protocol differs from Conventional OS.

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