

# Surface osteosarcomas: Diagnosis, treatment and outcome

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## ABSTRACT

Surface osteosarcomas are a rare form of osteosarcomas accounting for around 3-6% of all osteosarcomas. Three major groups of surface osteosarcomas are parosteal, periosteal and the high grade surface osteosarcomas. Of these, the parosteal osteosarcoma is the most common. Parosteal and periosteal osteosarcomas are distinct clinical entities and it is important to identify the clinicoradiological differences between the two types. Surface osteosarcomas occur at a later age as compared to conventional osteosarcomas. The classical site is the lower end of the femur followed by the upper end of the tibia and upper end of humerus, in that order. The periosteal variant affects the tibia more commonly than the parosteal variety. Neo-adjuvant chemotherapy is the standard of care for high grade surface osteosarcomas. Parosteal osteosarcomas, being low grade lesions, can be treated by upfront wide excision without adjuvant systemic therapy. Controversy prevails over the need for chemotherapy in periosteal osteosarcomas, which are intermediate grade lesions.

**Key words:** High grade, parosteal, periosteal, surface osteosarcoma

## INTRODUCTION

Surface osteosarcomas are rare primary bone tumors, which include a group of heterogeneous lesions with similar clinical, radiological, pathological and management end points. These tumors account for about 3-6% of all osteosarcomas.<sup>1</sup> The main variants of surface osteosarcomas are the parosteal, periosteal and the high grade surface osteosarcomas. Conventionally parosteal osteosarcomas are low grade tumors but dedifferentiated variants of parosteal osteosarcoma are not uncommon.<sup>2-4</sup> The high grade surface osteosarcomas have the worst prognosis. Various differences between parosteal, periosteal and high grade surface osteosarcomas are enumerated in Table 1. According to Grimer *et al.*, the main factors that differentiate a parosteal from a periosteal osteosarcoma is that a parosteal tumor arises on the surface of the bone

and has a high degree of structural differentiation, with a densely ossified mass radiologically and a low grade histological picture.<sup>5</sup> Periosteal osteosarcoma, on the other hand, arises from under the periosteum and the typical radiological feature is the periosteal elevation encircling a good proportion of the bone.

## PAROSTEAL OSTEOSARCOMA

The diagnostic criteria to designate a surface osteosarcoma as parosteal osteosarcoma were defined by Okada *et al.*<sup>6</sup> They concluded that radiographically, the lesion should have arisen from the surface of the bone, histologically, the tumor should be well differentiated (Grade 1 or 2); it should be characterized by well formed osteoid within a spindle-cell stroma and medullary involvement if any should be <25% of the medullary cavity.

## Demographics

Most studies on surface osteosarcomas are limited by small numbers, making it difficult to understand the actual demographics of the disease process. In a study by Song *et al.* a very high preponderance of female patients was seen.<sup>7</sup> There were 10 females in a group of 11 parosteal osteosarcomas in this retrospective study, treated over a period of 17 years. Most other studies show an equal propensity of surface osteosarcomas in both genders. Parosteal osteosarcomas show a mild female preponderance (male: female = 2:3). In one of the largest series of 226 parosteal osteosarcomas, the

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Access this article online	
Quick Response Code:	Website: www.ijonline.com
	DOI: 10.4103/0019-5413.132503

**Table 1: Differences amongst various subtypes of surface osteosarcomas**

Characteristic	Parosteal osteosarcoma	Periosteal osteosarcoma	High grade surface osteosarcoma
Grade	Usually low grade	Intermediate grade	High grade
Age of affection	Between 20 and 30 years	Between 18 and 20 years	Between 20 and 30 years
Gender	More common in females	Common in males (as compared to parosteal type)	Common in males
Site of affection	Posterior aspect of the distal femur is the commonest site	Metaphysio-diaphyseal portion of the tibia	Mid femur, distal femur, mid tibia (in order)
Radiological picture	Sclerotic lesion over the surface of bone with thickening of the cortex and presence of a periosteal line between the tumor and the normal bone (string sign)	Broad based soft tissue mass with destruction of underlying bone with perpendicular periosteal reaction going into the soft tissue mass	Lesion attached to the host bone with a broad base with dense to moderate mineralization with a fluffy, immature appearance noted prominently at the base of the lesion. Lucent zone and perpendicular periosteal reaction are absent
Histopathology	Characterized by being well differentiated with malignant osteoid formation and a spindle cell stroma. Most tumors are Grade 1 lesions	Shows lobulated islands of malignant cartilage and areas of moderately high grade spindle cells located peripherally	Shows areas of malignant spindle cells with a high degree of cellular atypia (Grade 3 or 4 lesion) and variable amount of osteoid formation
Treatment	Minimal role of neo-adjuvant chemotherapy. Upfront surgery is the standard of care. Dedifferentiated tumors are high grade lesions that need to be treated like conventional osteosarcoma	Role of chemotherapy is controversial. Wide excision is the preferred local therapy	Neo-adjuvant chemotherapy followed by surgery

average age of patients was 28 years (range 8-64 years).<sup>6</sup> In a review of 29 cases of dedifferentiated parosteal osteosarcomas from the Rizzoli Institute, the average age was 36 years (range 15-85 years).<sup>2</sup> It is therefore clear that the mean age of surface osteosarcomas is high when compared to conventional osteosarcomas, but the exact period of the decade when these occur is variable in most series. Dedifferentiated parosteal osteosarcomas, which are high grade lesions, can occur in a wide age distribution.

### Histopathology

Histological grade of the lesion is essential to classify surface osteosarcomas. The commonly used four tier grading system is based on the original Broder's grading.<sup>8</sup> The numeric grade of the tumor from 1 to 4 was equated to the percentage of anaplasia within the tumor (from  $\leq 25\%$  to 100%). Parosteal osteosarcomas are usually Grade 1/Grade 2 osteosarcomas that either lack or have minimal anaplasia and it is difficult to recognize these entities as neoplasms on cytological basis alone. Grossly, they appear as hard lobulated mass attached to the underlying cortex<sup>9</sup> [Figure 1]. Nodules of cartilage may be present within the substance of the tumor, or there can be an incomplete cartilage cap at the surface. Microscopic examination will demonstrate parallel, well formed bony trabeculae in a hypocellular stroma with or without osteoblastic rimming [Figure 2]. Should a focus of anaplasia is identified within what is otherwise a Grade 1 tumor, then the lesion is termed "dedifferentiated" and it is assumed to follow an aggressive course. Dedifferentiated parosteal osteosarcomas can be either synchronous (diagnosed at presentation) or metachronous (diagnosed at the time of recurrence). Cytogenetic analysis of parosteal osteosarcoma has demonstrated gain of 12q13-15 sequence contained within supernumerary ring chromosomes.<sup>9,10</sup>

This characteristic cytogenetic abnormality is uncommon in conventional osteosarcoma. Such ring chromosomes are noted in other low grade malignant mesenchymal neoplasms such as well differentiated liposarcoma and dermatofibrosarcoma protuberans.<sup>1</sup> The loci for SAS gene is located in q13-15 region of chromosome 12 and this gene is found to be amplified in surface osteosarcoma.<sup>11</sup> Other genes, which were frequently found to be either co-amplified or over expressed in parosteal osteosarcoma include CDK4 and MDM2.<sup>12</sup>

### Clinical presentation

The most common location of a parosteal osteosarcoma is the posterior and distal part of the femoral metaphysis [Figure 1]. This is followed by the proximal tibial metaphysis and the proximal humeral metaphysis. These three locations accounted for >80% lesions in the Okada *et al.* series.<sup>6</sup> In contrast, dedifferentiated parosteal osteosarcomas involve long bones (femur, humerus and tibia in order) as was seen in a series from the Rizzoli Institute.<sup>2</sup>

The clinical presentation is varied with most patients coming late for consultation in view of the lesion being slow growing. The commonest presentation is pain and swelling with >80% of the lesions being around the knee joint. In Okada's *et al.* series of 189 parosteal osteosarcomas, 80 patients were painless.<sup>6</sup> Swelling was the most common feature in this series. Swelling was followed by pain as the second common feature in their series, seen in 66 patients. 33% of the patients in this series had limitation of knee movements.

Systemic metastasis is rare in parosteal osteosarcomas. In Okada *et al.* series, none of the patients had metastasis at presentation.<sup>6</sup> Song *et al.* reviewed the records of 22

parosteal osteosarcoma patients and none of them had metastatic disease at presentation.<sup>13</sup> In a series of 63 cases of parosteal osteosarcoma treated between 1978 and 2007, Zaikova *et al.* reported 9 (14.3%) patients developing metastases at a median of 22 months (6-123 months).<sup>14</sup> Five patients developed both locoregional and metastases. Seven patients in this series died from the tumor, while two patients were alive with metastatic disease.

### Imaging

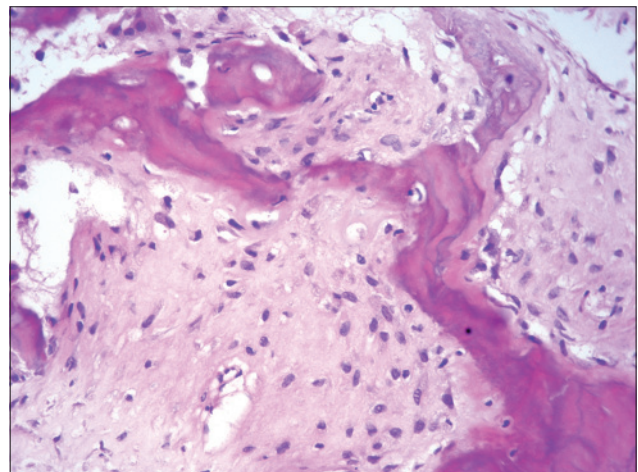
The radiological picture of surface osteosarcomas is characteristic. Campanacci *et al.* had described parosteal osteosarcomas as "...lesions projecting from the periosteal tissues with a mushroom-like radiographic appearance".<sup>15</sup> On plain X-rays parosteal osteosarcoma appears as a heavily mineralized mass attached to the cortex with a broad base. In their study Okada *et al.* they found that most tumors (70%) had a cortical attachment of 1 cm or more.<sup>6</sup> The tumor has a tendency to wrap around the involved bone as it grows. Around 80% tumors involved either half or less than half the circumference of bone. The underlying cortex can be normal, thickened, or destroyed. A thin lucent zone between the tumor and the host bone was noted in >50% cases of parosteal osteosarcoma. However, this lucent zone was more evident on computerized tomography (CT) rather than on a radiograph [Figure 2]. Periosteal new bone formation and medullary involvement are extremely rare (<10%). Lucent areas within the substance of tumor, as noted in plain radiograph and CT scan, could indicate the presence of dedifferentiated areas in parosteal osteosarcoma.<sup>2</sup> Positron emission tomography-CT scans can be a valuable tool in the evaluation of such lesions.<sup>16</sup> They can demonstrate areas of high metabolic activity inside the lesion that can be specifically targeted and biopsied.

### Treatment

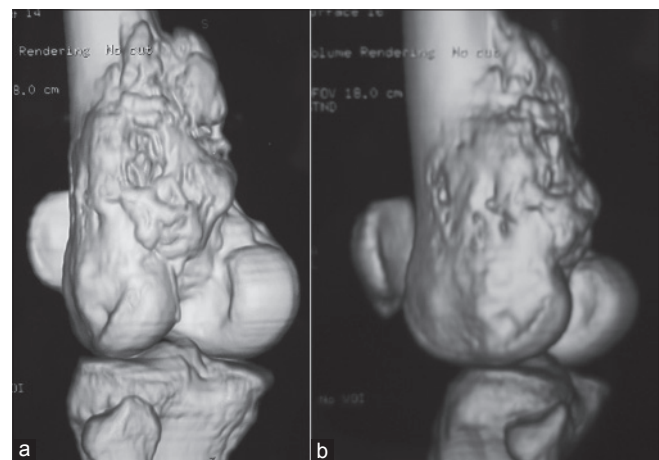
All surface osteosarcomas need surgical excision regardless of the grade of the lesion. Parosteal osteosarcomas, being low grade lesions, can be treated by upfront wide excision without adjuvant chemotherapy and they require either marginal or wide excision. The technique of hemicortical resection for treating parosteal osteosarcomas was first described by Campanacci *et al.* in 1982 [Figure 4].<sup>17</sup> The decision on whether a lesion can be addressed by hemicortical excision should be taken after carefully evaluating the preoperative imaging modalities and histopathological grade. Only those lesions not involving the neuro vascular bundle in which surgical excision would not include the adjacent articular surface and at least one third of the bone's circumference would remain after excision should undergo hemicortical resection.<sup>18</sup> The technique involves marking the line of resection around the base of tumor taking surgical margin



**Figure 1:** Plain radiograph of the knee (lateral view) in a 26 year old lady with a parosteal osteosarcoma of the posterior surface of distal femur (commonest location)



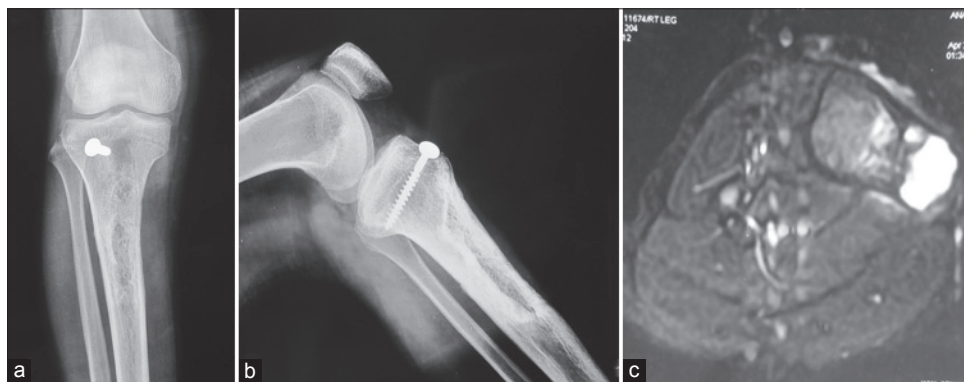
**Figure 2:** A photomicrograph (×20) of a parosteal osteosarcoma showing well formed bony trabeculae in a hypocellular stroma with or without osteoblastic rimming



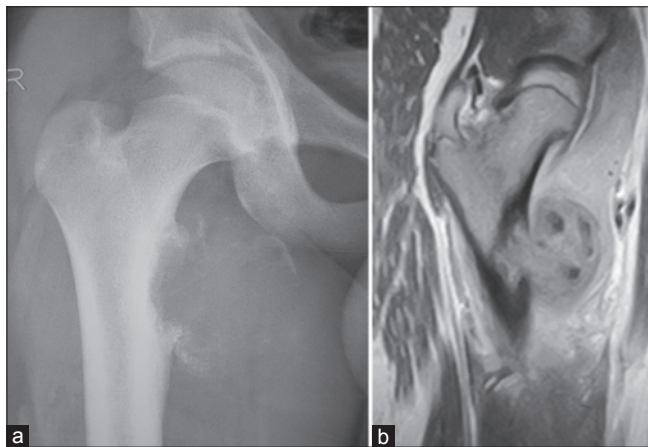
**Figure 3:** (a and b) 3-D CT reconstruction of a parosteal osteosarcoma of the distal third of the femur in a 24-year-old male patient



**Figure 4A:** (a) Clinical photograph of a 20-year-old male patient showing parosteal osteosarcoma of the right proximal tibia (b) Per-operative pictures of the same patient showing hemicortical resection of the anterior tibia (note the K wire holding the patellar tendon) (c) Same patient showing wound closure over a gastrocnemius flap (d and e) At 4 years followup showing healed scar and range of motion



**Figure 4B:** (a and b) Plain radiographs (anteroposterior and lateral views) of the leg (followup after 3 years) showing good consolidation of the hemicorticotomy (c) MRI (T2 weighted image) of the tibia showing mainly a hyperintense surface mass with minimal involvement of the medullary cavity



**Figure 5:** (a) Plain radiograph anteroposterior view of the upper right femur with hip joint in a 14 year old boy showing a destructive expansive lesion arising from the medial cortex of a upper femur. Histologically it was a periosteal osteosarcoma (b) MRI of the same patient showing a huge soft tissue component of the surface lesion. Typical of a periosteal osteosarcoma

into account and creating a unicortical window along this line with the help of a saw. The defect can be reconstructed using either bone cement, autograft (e.g. fibular autograft), allograft or pasteurized/autoclaved/irradiated host bone

[Figure 4A and B]. Song *et al.* retrospectively reviewed the results of 22 parosteal osteosarcoma patients treated by hemicortical excision and reconstruction.<sup>13</sup> The 10-year overall survival was 85.7% with an event free survival of 54.5%. Seven of their patients underwent intralesional excision due to mis-diagnosis and all of them developed local recurrence. Two of the 5 patients who underwent marginal excision developed local recurrence of which one lesion was dedifferentiated, whereas the other was histological Grade 2. None of their patients developed local recurrence after wide excision. Zaikova *et al.*, in their review of 63 patients, reported a local recurrence rate of 46% following intra-lesional excision as compared to 20% following marginal excision and 0% following wide excision.<sup>14</sup> Wide margin following hemicortical resection was also reported by Liu *et al.*<sup>18</sup> None of their patients developed local recurrence. Thus, we find that en-bloc resection with wide surgical margins is the gold standard, while hemicortical excision has a role in selected cases of parosteal osteosarcoma.

### Differential diagnoses

Surface osteosarcomas can mimic benign conditions such as osteochondroma and myositis ossificans. Song *et al.* in their

study have reported that five cases of parosteal osteosarcomas that were treated with intralesional therapy mistaking them for benign conditions and all of these presented with local recurrence.<sup>7</sup> Diagnosing these tumors need a high index of suspicion. All clinicoradiologically doubtful lesion needs to be biopsied to confirm the diagnosis. Cartilage cap of parosteal osteosarcoma, if present, lacks the columnar arrangement of cells and shows mild cellular atypia thus differentiating it from osteochondroma. Difficulty in diagnosing these lesions gets compounded when they present at unusual locations. For example, parosteal osteosarcoma arising from the bones of hand need to be differentiated from Nora's lesion (Bizarre parosteal osteochondromatous proliferation) and Turret's exostosis.<sup>19</sup>

## PERIOSTEAL OSTEOSARCOMA

Periosteal osteosarcoma is an intermediate grade chondroblastic osteosarcoma arising on the surface of the bone.<sup>9</sup>

### Demographics

Most surface osteosarcomas have a slightly higher age of occurrence when compared to conventional osteosarcomas. However, in a study including 40 patients of periosteal osteosarcoma, the mean age of patients was 20 years (range 10-37 years).<sup>20</sup> Thus, amongst these surface lesions, the periosteal subtype tends to occur at a younger age, followed by parosteal and high grade surface osteosarcomas, which are more common during the third decade. Periosteal osteosarcomas are more common in males.

### Histopathology

Unni *et al.* first described periosteal osteosarcoma, as a distinct entity.<sup>21</sup> The lesion is characterized by lobulated islands of malignant cartilage and areas of moderately high-grade spindle cells located peripherally. Trabeculae of mature osteoid are absent and the lesion shows little tendency to invade skeletal muscles.

### Clinical presentation

Pain and swelling are the usual presenting symptoms. However, periosteal osteosarcoma is considered to be more painful and runs a rapid course when compared to parosteal osteosarcoma. This subtype of surface osteosarcoma affects tibia more commonly than femur and the location is typically metadiaphyseal.<sup>21</sup> The tibial lesions are more common in the proximal third, whereas the femoral lesions are more common in the middle and distal third. Periosteal osteosarcomas resemble parosteal variety in having lower incidence of systemic metastasis. In a series of 119 patients of periosteal osteosarcoma, Grimer *et al.* noted metastasis in 17 patients (14%), a previous local recurrence ( $n = 4$ ), lung metastases ( $n = 16$ ) and bone metastasis in the opposite leg

( $n = 1$ ). Two patients had both lung and bone recurrences in this study.<sup>5</sup>

### Imaging

Periosteal osteosarcomas are surface lesions with nonhomogenous calcified spiculations perpendicular to the cortex giving a "sunburst appearance".<sup>9</sup> The lesion decreases in density from the cortical base to the surface. A radiological review of 40 cases of periosteal osteosarcoma found that the lesion usually appears as a broad based soft tissue mass causing extrinsic erosion of thickened underlying diaphyseal cortex and perpendicular periosteal reaction extending into the soft tissue component [Figure 5].<sup>20</sup> According to Murphey *et al.*, reactive marrow changes are common but true marrow invasion is rare.<sup>20</sup> However, certain other authors report high (20-70%) incidence of medullary invasion.<sup>22,23</sup>

### Treatment

Periosteal osteosarcomas should be treated with en-bloc resection with wide margins based on established oncological principles as with conventional osteosarcomas. Controversy prevails over the need for neo-adjuvant chemotherapy in periosteal osteosarcomas, which are intermediate grade lesions. Revell *et al.* retrospectively reviewed 17 cases of periosteal osteosarcoma at a mean followup of 52 months.<sup>23</sup> Patients in their study were given chemotherapy if tumor showed high grade histological features or if medullary involvement was present in magnetic resonance imaging (MRI). Ten of their patients received neo-adjuvant chemotherapy, 4 received adjuvant chemotherapy while 3 patients did not receive any adjuvant therapy. One patient with positive surgical margin developed local recurrence while the survival rate at 52 months was 100%. They advocated neo-adjuvant chemotherapy for all periosteal osteosarcoma patients with high areas or medullary involvement. Cesari *et al.* reported their experience of treating 33 periosteal osteosarcoma patients.<sup>22</sup> Nineteen patients did not receive any adjuvant treatment, 14 received adjuvant chemotherapy and 4 received neo-adjuvant chemotherapy. They did not report any criteria used to decide on whether to administer chemotherapy or not and they did not follow fixed chemotherapy protocol in within any group. All 14 patients subjected to chemotherapy had Grade 3 tumors. The 10 year overall survival rate was 86% in the chemotherapy group and 83% in the only local therapy group. They concluded that adjuvant chemotherapy did not improve survival. Grimer *et al.* reviewed the effect of chemotherapy on the outcome of periosteal osteosarcoma patients. This was a multicenter review from the European Musculo Skeletal Oncology collaboration.<sup>5</sup> No criteria were followed, while deciding whether or not to administer chemotherapy. The protocol also differed among different centers. They could not demonstrate the use of chemotherapy as a prognostic

factor for periosteal osteosarcomas. However, survival was related to appearance of local recurrence ( $P < 0.0001$ ) and none of patients with  $>90\%$  necrosis following neo-adjuvant chemotherapy developed local recurrence. Given the uncertainty regarding the benefit of chemotherapy and the lack of universally accepted chemotherapy protocols, only a randomized controlled trial (RCT) can provide an answer. However, periosteal osteosarcoma, being a rare disease, is not an ideal candidate for a RCT.<sup>23</sup> The authors opine that the decision on whether to administer chemotherapy or not has to be taken at the individual tumor board taking into consideration the local factors affecting such decision. In our institute, we prefer neo-adjuvant chemotherapy in most periosteal osteosarcoma patients. The rationale is that we work in a resource constrained environment where a delay in surgery is expected due to long waiting list and the patient receives neo-adjuvant chemotherapy, while awaiting his/her surgical slot.

### Differential diagnosis

Periosteal osteosarcomas need differentiation from conventional osteosarcomas, which are high grade tumors and periosteal chondroma, a benign cartilaginous tumor. Confusion prevails over the term “juxtacortical chondrosarcoma”, supposedly the malignant counterpart of periosteal chondroma. Some authors classify juxtacortical chondrosarcoma and periosteal osteosarcoma as different entities, while others use juxtacortical chondrosarcoma as a synonym for periosteal osteosarcoma, thus confirming that both the lesions are one and the same.<sup>9,24</sup>

## HIGH GRADE SURFACE OSTEOSARCOMA

According to Staals *et al.*, the first description of high grade surface osteosarcoma was by Francis in 1964.<sup>25</sup> As the name suggests, high grade surface osteosarcomas are highly malignant lesions that has the ability to metastasize and cause death similar to conventional osteosarcoma.

### Demographics

Majority of the studies on surface osteosarcomas focus only on low grade parosteal variety, thus limiting our understanding on the other surface osteosarcomas. In a study on 46 patients of high grade surface osteosarcoma, the mean age was found to be 25 years (range 8-70 years).<sup>26</sup> In another study from Rizzoli Institute, there were 19 males and 6 females with a mean age of 21 years.<sup>25</sup> High grade surface osteosarcomas, like periosteal osteosarcomas, are commoner in males.<sup>27</sup>

### Histopathology

The histology of high grade surface osteosarcomas demonstrates areas of spindle cells with cellular atypia (Broder's Grade 3 or 4) and varied amounts

of osteoid formation.<sup>8</sup> Grade 3 tumors lack cellular pleomorphism (marked variation in size and shape of nuclei) as opposed to Grade 4 tumors that show marked pleomorphism. The most common histologic subtype was osteoblastic followed by chondroblastic subtype.<sup>25</sup>

### Clinical presentation

High grade surface osteosarcomas follow an aggressive clinical course. They most frequently involved the midfemur followed by distal femur and midtibia.<sup>26</sup> Pain and swelling are the usual presenting symptoms.

### Imaging

High grade surface osteosarcomas present a similar radiological picture as other surface osteosarcomas [Figure 3]. Okada *et al.* have reported several radiological features that might aid identification of these lesions.<sup>26</sup> Most high grade surface osteosarcomas presented dense to moderate mineralization with a fluffy, immature appearance which was more prominent at the base of the lesion. The lesions were attached to the host bone with a broad base and a lucent zone between the lesion and host bone was rare as was circumferential bony involvement. In contrast to periosteal osteosarcomas, spiculated periosteal reaction perpendicular to host bone was rarely encountered in high grade surface osteosarcomas.

### Treatment

Neo-adjuvant chemotherapy is the standard of care for high grade surface osteosarcomas. The chemotherapy protocol is similar to conventional osteosarcoma and all lesions should undergo excision with wide oncological margins. Metastatic lesions need to be treated in a similar way as conventional osteosarcoma. In the Rizzoli Institute study, the 5-year overall survival was 82% and disease free survival was 70%.<sup>25</sup>

### Differential diagnosis

High grade surface osteosarcomas need to be differentiated from the parosteal and periosteal subtypes as the treatment principles differ. The other differentials include extra skeletal osteosarcoma and periosteal Ewing's sarcoma without medullary involvement.

## CONCLUSION

Surface osteosarcomas are distinct clinicopathological entities. Their recognition is important as the prognosis and treatment protocols differ significantly from conventional osteosarcoma. Radiological tools such as CT scan and MRI are vital to the diagnosis and surgical planning of these lesions. Although all of them are neoplastic lesions, the malignant potential varies significantly with parosteal

osteosarcoma being the least malignant followed by periosteal and high grade surface subtypes in that order. Dedifferentiated parosteal osteosarcoma is a high grade lesion and its management is similar to high grade surface osteosarcoma. Parosteal osteosarcoma can be treated by upfront surgery without neo-adjuvant chemotherapy, while high grade lesions should be subjected to neo-adjuvant protocol similar to conventional osteosarcoma. The role of neo-adjuvant chemotherapy in periosteal osteosarcoma is controversial. Overall, surface osteosarcomas carry either similar or better prognosis when compared to conventional osteosarcomas depending upon the subtype and hence, the need to identify the different subtypes cannot be over emphasized.

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**How to cite this article:** Kumar VS, Barwar N, Khan SA. Surface osteosarcomas: Diagnosis, treatment and outcome. *Indian J Orthop* 2014;48:255-61.

**Source of Support:** Nil, **Conflict of Interest:** None.