The tumor appears as a hard lobulated mass attached to the underlying external layer of periosteum, and may contain nodules of cartilage within the substance of the tumor or an incomplete cartilage cap at the surface [4]. Microscopically, PO is characterized by hyalinized fibrous stroma with a low cell content without substantial nucleus polymorphism and variably dense bony trabeculae [3]. In some cases, the diagnosis of PO can be complicated by the presence of highly differentiated areas with fatty tissue within the marrow and uniform bony structure [3].

Here, we report a case of focal fatty metaplasia identified within a PO lesion. Given a lack of previous reports on this rare condition, we offer a characterization of this rare histomorphological variation to enrich the morphological spectrum of PO.

**Case Description**

A 28-year-old man was referred to our orthopedic department surgery for a mass that was identified in her right tibia on radiography after sudden injury of her right lower limb 2 months prior. The patient did not experience any pain or swelling related to the mass. A biopsy performed at another hospital did not reveal any histologic evidence of malignancy. On physical examination, the patient presented with a hard, poorly circumscribed, painless, and immobile palpable mass at the posterior aspect of the proximal right tibia. The area surrounding the mass was slightly warm, but

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**INTRODUCTION**

Parosteal osteosarcoma (PO) is a typically well-differentiated subtype of osteosarcoma, with slow growth and low grade malignancy that do not tend to metastasize. PO is the most common type of osteosarcoma arising on the bone periosteum and accounts for 2%-5% of all primary malignant bone tumors, approximately 6%-8% of all osteosarcomas, and 75% of surface osteosarcomas [1].

PO mainly affects adults between the ages of 18-35 years and exhibits a higher prevalence in women. The most common sites affected by PO include the knee, accounting for about 75% of cases [2] and specifically the posterior aspect of the distal femur, followed by the proximal tibia, proximal humerus; only 6% of all POs are observed in the skull, the spine, and pelvis [3].

The radiographic feature of osteosarcoma includes a lobulated exophytic mass with central dense ossification adjacent to the bone usually in the posterior aspect of the distal femur, sparing the medullary canal.

Tomography scans reveal well-defined, hyperdense bony mass with patchy radiopacification.

PO originates from the outer fibrous layer of the periosteum and is mainly composed of fibroblasts.

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**Case Report**

It is the most common type of juxtacortical or surface osteosarcoma and accounts for 5% of all osteosarcomas. It typically presents in early adulthood and middle age with a peak incidence in the third decade. It affects females slightly more than males. Patients usually present with a painless, slowly enlarging mass. They are usually located at the metaphysis (80%-90%), most commonly at the posterior aspect of the distal femur (60%), followed by either end of the tibia, and then the proximal humerus. Parosteal osteosarcomas are usually low-grade lesions are usually treated with surgical resection and no neoadjuvant chemotherapy or radiation. As they are frequently metaphyseal in location, large parosteal osteosarcomas or those with deep medullary invasion may require limb salvage, including joint replacement. Parosteal osteosarcomas have an excellent prognosis (80-95% long-term survival).

**Keywords:** osteosarcoma; adult, metaphysis, low grade, surgery.
no changes in the skin were noted. The patient had normal range of motion of the right knee and no abnormal findings on hematologic or biochemical studies.

Radiography of the knee revealed a well circumscribed bone forming lesion associated with the external periosteum of the distal femur (Fig-1).

Computed tomography showed no medullary involvement and confirmed that the bony mass arose from the periosteum, exhibiting a lower density than the femur cortex. The femur cortex appeared to be intact. All findings were compatible with PO. Additionally, 2 areas of fatty density were observed in the osteoid tumor.

Fig-1: The lateral view of the radiography revealed an ossified mass on the periosteum (arrowheads) with a presence of string sign, a radiolucent line separating the ossified mass and underlying cortex (arrows); MRI of the sagittal section of the mass revealed a heterogeneous signal, with the solid component predominately hypointense on T1-WI (arrow) (1B), hyperintense on T2-WI (arrow) (1C), and well enhancement on T1-WI with fat saturation after gadolinium contrast enhancement (arrow) (1D)

The patient subsequently underwent resection of the affected segment of the distal right femur and reconstruction of the right knee and femur by prosthesis implantation. The hematoxylin and eosin stained histologic diagnosis was PO with focal fatty metaplasia (Fig. 3). Good histologic margins were achieved and there was no tumor recurrence or metastasis after the resection.

Okada et al., [5] defined criteria for the radiological diagnosis of PO to include lesions arising from the surface of the bone with good differentiation of the tumor on histology (grade 1 or 2); a well formed osteoid within a spindle cell stroma; and, in cases of medullary involvement, occupation of < 25% of the medullary cavity. A thin radiolucent zone is seen between the tumor mass and host bone cortex in > 50% cases, and the cortex of the host bone can be normal, thickened, or destroyed.

Cytogenetic analysis of PO has demonstrated supernumerary ring chromosomes containing gain of 12q13-15 sequence.
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 [7]. SAS gene is located in q13-15 region of chromosome 12 and is found to be amplified in surface osteosarcomas, also in the parosteal subtype [8].

The differential diagnosis of PO usually includes some benign conditions such as osteochondroma and myositis ossificans; the first usually presents corticomedullary continuity with the underlying medullary canal, while myositis ossificans presents a peripheral ossification, inverse of the PO ossification pattern, with calcifications centrally located.

Diagnosing a PO needs a high index of suspicion. All clinicoradiologically doubtful lesion needs to be biopsied to confirm the diagnosis. The natural history of PO is relatively benign. Its optimal treatment is wide surgical resection, with an overall survival rate of 85%-92%. Incomplete removal is associated with local recurrence and increased risk of metastasis [5]. In the present case, we observed radiolucent areas in the tumor that were later identified on histology as focal fatty metaplasia. Because of the less aggressive biological behavior, it is important to know this variant of parosteal osteosarcoma, in order to recognize and differentiate it from benign osseous lesions.

**Fig. 2:** Sheets of metaplastic adipocytes among the spindle tumor cells of a parosteal osteosarcoma (H&E stain, ×100)

**REFERENCES**