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Background: Osteoblastoma is a benign osteoblastic neoplasm with an aggressive growth pattern. This tumor constitutes about 1% of excised primary bone tumors. Pain is the most common symptom. Most patients are between 10 and 30 years of age. Osteoblastoma most commonly occurs in the spine, followed by the diaphysis and metaphysis of long bones. It rarely arises from the periosteum. Histologically, osteoblastoma consists of well-vascularized connective tissue stroma with interconnecting trabeculae of immature woven bone. Epithelioid osteoblastomas constitute an even rarer subset of osteoblastomas characterized by a locally aggressive, relentless growth pattern and histology that may be extremely difficult to distinguish from an osteosarcoma.

Question/ Purpose: In this paper, we report a rare case of a giant periosteal osteoblastoma in the femur of a 21-year-old male. The goal of this study is to aid professionals in the diagnosis and treatment of highly aggressive osteoblastomas.

Patients and Methods: A 21-year-old male presented with a painful, very firm, nontender mass from his left thigh. The pain was worse at night and was relieved with NSAIDs. He had no fevers, night sweats, or weight loss. The patient underwent preoperative radiological studies including plain radiographs, MRI, and CT scan. An open biopsy was performed that was consistent with an aggressive, epithelioid osteoblastoma. The patient subsequently underwent resection and cryosurgery with fixation and bone grafting with cortical strut allografts.

Results: Pathology demonstrated a neoplasm characterized by cohesive sheets of epithelioid osteoblasts, mixed with areas of conventional osteoblastoma displaying prominent osteoblastic rimming of woven bone trabeculae in a fibrovascular stroma. Preoperative X-rays demonstrated diffuse cortical thickening along the distal diaphysis of the femur. It was difficult to discern any lesion. MRI confirmed diffuse cortical thickening and absence of involvement of the medullary canal except by surrounding edema. CT scan was the key study for identifying the lesion. This surface based tumor was well circumscribed, heavily mineralized, and surrounded by an extensive benign appearing periosteal reaction. The medullary canal appeared free of neoplasm. The patient underwent resection of tumor of the left femur, cryosurgery, and fixation of the left femoral shaft and bone grafting of the posterior cortex with cortical strut allografts. One year postoperatively, radiographs demonstrated solid healing at the allograft site. At final follow-up, 32 months postoperatively, there was no evidence of local recurrence. The patient had resumed all his normal activities. He could run without pain and had no restrictions with activities.

Conclusions: Aggressive periosteal epithelioid osteoblastoma is an extremely rare entity. Radiological-pathological correlation is crucial for proper diagnosis and distinguishing it from an osteosarcoma, particularly an osteoblastoma-like osteosarcoma. This tumor is locally destructive and causes significant pain. Historically, aggressive osteoblastomas have had a high incidence of local recurrence. Surgery is the mainstay of treatment. Adjuvants such as cryosurgery may be considered to help eradicate microscopic disease, decrease local recurrence and preserve as much bone as possible for reconstruction and to avoid wide en bloc resection. Even though most local recurrences of benign aggressive tumors occur within two years of surgery, further long term follow up of this patient is required especially given the paucity of information about this rare entity.