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Case Study

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FEMORAL PAROSTEAL OSTEOSARCOMA: CASE REPORT

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ABSTRACT

Introduction: parosteal osteosarcoma is a low-grade, bone-forming neoplasm that arises on the surface of bone. It accounts for about 4% of all osteosarcomas. **Case report:** an 18-year-old male patient with no particular past medical history, consulted for a painless mass in the right thigh that had appeared at the age of 17 years and progressively increased in volume. The physical examination revealed a 6 cm mass at its largest above the right popliteal fossa with knee flexion slightly limited. The X-ray revealed a well-limited mass in the lower third of the femur that was dense and attached to the metaphyseal cortex by a wide base. Histological examination of the biopsy specimen established the diagnosis of parosteal osteosarcoma. The patient underwent wide resection of the femoral tumor preceded by a course of first-line chemotherapy. Postoperative course was uneventful. During the one-year follow-up period, there was no recurrence or metastasis of the tumor. **Conclusion:** parosteal osteosarcoma is characterized by its insidious growth and favorable prognosis. It rarely leads to metastasis. Its treatment is mainly surgical.

KEYWORDS: Parosteal osteosarcoma, bone, tumor, pathology.

INTRODUCTION

Parosteal osteosarcoma, or juxtacortical osteosarcoma, is a rare subtype of osteosarcoma that develops at the bone surface, but has a more favorable prognosis than other conventional osteosarcomas.[1] In this article, we report a new case of parosteal osteosarcoma in an 18-year-old male patient whose initial presentation was a swelling of the right thigh. Our aim was to review the clinical, radiologic, and pathologic features of parosteal osteosarcoma, with a special emphasis on the differential diagnosis and the recent advances in molecular biology.

CLINICAL HISTORY

An 18-year-old male patient with no particular past medical history, consulted for a painless mass in the right thigh that had appeared at the age of 17 years and progressively increased in volume. The physical examination revealed a 6 cm mass at its largest above the right popliteal fossa with knee flexion slightly limited. The X-ray revealed a well-limited mass in the lower third of the femur that was dense and attached to the metaphyseal cortex by a wide base. Histological examination of the biopsy specimen demonstrated a moderately cellular malignant tumor proliferation, with moderately atypical and fusiform cells surrounded by osteoid trabeculae. Mitotic figures were infrequent. The preoperative diagnosis was that of parosteal osteosarcoma. The patient underwent wide resection of the femoral tumor preceded by a course of first-line chemotherapy. The macroscopic examination of the specimen showed a 6.3 cm×2.8 cm tumor of the distal femur. It was located 3 cm from the limit of surgical resection. The cut specimen presented a lobulated, whitish aspect with focal cartilaginous zones (Figure 1). There was no tumor invasion of the skeletal muscle and the surgical limits. The histological analysis showed a malignant mesenchymal proliferation, moderately cellular, made up of long, linear and eosinophilic material, sometimes calcified with no osteoblastic cells in the periphery (Figure 2). The tumor cells were spindle-shaped, with little eosinophil cytoplasm and a long or ovoid, hyperchromatic, and moderately atypical nucleus (Figure 3). Mitoses were rare. There were no areas of dedifferentiation. The final pathological diagnosis was that of parosteal osteosarcoma with no dedifferentiation areas, with healthy excision margins. Postoperative course was uneventful. During the one-year follow-up period, there was no recurrence or metastasis of the tumor.

FIGURE LEGENDS

Figure 1: Macroscopic examination of the surgical specimen showing a lobulated, whitish tumor with focal cartilaginous zones measuring 6.3 cm x 2.8 cm, attached to the bone by a wide base.

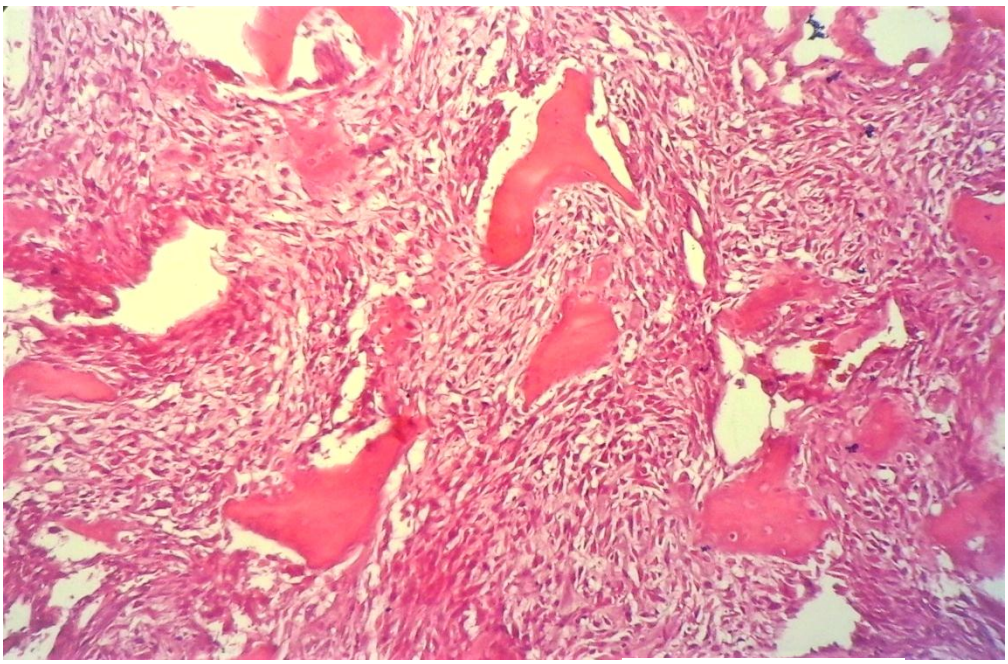

Figure 2: Photomicrograph of parosteal osteosarcoma showing mature-appearing bone without osteoblastic rimming, surrounded by a hypercellular fibroblastic stroma with moderate cytologic atypia, magnification (×200).

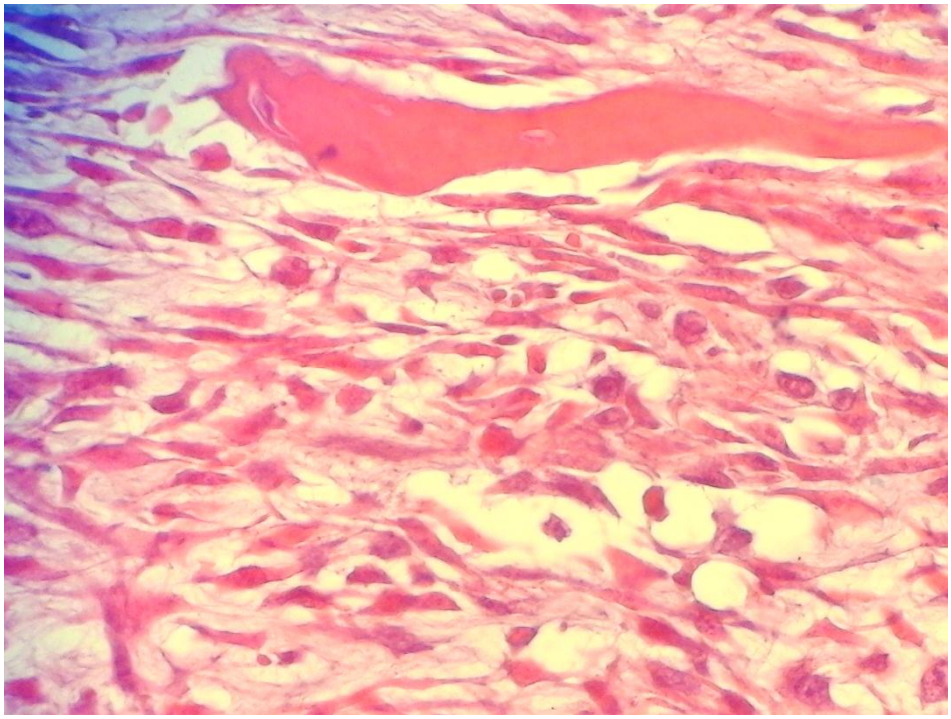

Figure 3: Tumor cells are spindle-shaped with a long or ovoid nucleus, hyperchromatic, and moderately atypical, magnification (×400).

DISCUSSION

Parosteal osteosarcoma is a low-grade, bone-forming neoplasm that arises on the surface of bone. It accounts for about 4% of all osteosarcomas and for more than 75% of all surface osteosarcomas. The peak age of presentation is in the second or third decade with a female predominance.[2,3] The most common location is the posterior aspect of the distal femur in approximately 70% of cases, followed by the proximal tibia and proximal humerus. Rare locations, including cranial, mandible, rib, clavicle, and tarsal bone, have been reported.[2,3] Patients with parosteal osteosarcoma present with a slowly growing painless mass, with decreased range of movement of the adjacent joint. Dull pain and tenderness are the second most common symptoms.[4] The protracted clinical course of parosteal osteosarcoma is an important feature that distinguishes this tumor from other benign lesions of similar locations such as myositis ossificans and high-grade surface osteosarcoma, which usually have a more rapid onset. Early diagnosis depends on clinical suspicion, meticulous radiologic evaluation, and accurate histologic analysis.[5] On plain radiograph, parosteal osteosarcoma usually presents as a lobulated, mushroom like tumor protruding from the underlying cortex with a broad base attachment. The periphery of the tumor is generally less radiodense than the center and the pattern of mineralization is irregular. Most of the tumors involve the metaphysis of the long bones while involvement of the diaphysis is rare.[6] Computed tomography scan defines the extent of the tumors for surgical planning, but cannot differentiate the lucent areas within dense tumors, which contain either benign tissue or tumor of any grade. Moreover, it may not reveal small satellite nodules beyond the main tumor.[7] Computed tomography scan is optimal for assessment of cortical integrity. Magnetic resonance imaging is the most appropriate imaging study to evaluate medullary involvement, satellite nodules and can guide the surgeons to ensure adequate resection of bone marrow infiltrated by tumor.[8] Grossly parosteal osteosarcoma presents as a hard lobulated mass, affixed to the underlying cortex, attached to the bone by a wide base, pushing the adjacent structures back. The cut surface has a heterogeneous aspect, showing fibrous zones and some cartilaginous zones. Intramedullary extension is seen in 25% of cases and must be searched for carefully in multiple samples.[6] The presence of soft and fleshy areas suggest dedifferentiation in contrast to the bony tumor. Focal necrosis and hemorrhage can be seen.[6] In our case, there were no zones of soft and fleshy consistency. Microscopically, parosteal osteosarcomas have a biphasic appearance. The tumor consists of spindle cells with minimal atypia, forming well-formed, bony trabeculae that are arranged in a parallel manner. Osteoblastic rimming may be present or absent in the trabeculae. The tumor tends to be hypocellular, although in about 20% of the cases, it is hypercellular and the spindle cells show moderate atypia. About 50% of the tumors show cartilaginous differentiation. This may be in the form of hypercellular nodules of cartilage within the substance of the neoplasm or as a cap on the surface. If present, the cartilage cap shows mild hypercellularity and the cells with mild cytological atypia and lack the ‘’columnar’’ arrangement seen in osteochondromas. Around 15-25% of the tumors will show high-grade spindle cell areas, indicating progression to high-grade sarcoma (dedifferentiation).[6] In the case reported herein,

no areas of dedifferentiation were found and the tumor was low grade (conventional parosteal osteosarcoma).

The differential diagnosis of low-grade parosteal osteosarcoma includes juxtacortical heterotopic ossification/myositis ossificans and osteochondroma. Corticomedullary continuity between the osteochondroma and the underlying bone is a characteristic feature present in osteochondromas. Parosteal osteosarcoma is devoid of corticomedullary continuity. Moreover, in the intertrabecular spaces of parosteal osteosarcoma there are neoplastic spindle cells rather than normal marrow elements. In myositis ossificans, there is exuberant heterotopic ossification on the periosteal bone surface or adjacent soft tissues that can overlap with parosteal osteosarcoma.[6] Recent studies demonstrated characteristic cytogenetic abnormality resulting in amplification of the CDK4 and MDM2 genes, which may serve as markers for molecular diagnosis.[9] In contrast to heterotopic ossification and osteochondromas, parosteal osteosarcomas show MDM2 amplification, which can be a very useful tool in difficult cases.[9-11] Surgery remains the treatment of choice for parosteal osteosarcoma. The most adequate treatment is wide excision of the tumor with more than a 1-cm surgical margin. However, incomplete excision almost inevitably leads to local recurrence.[12] In case of recurrence, re- amputation or excision may provide a possible cure in cases that lack tumor dedifferentiation.[13] A preoperative diagnosis and radiological assessment of the extent of the tumor are required in order to achieve complete excision. Parosteal osteosarcomas can undergo dedifferentiation to a high-grade osteosarcoma. This phenomenon occurs in an estimated 15 to 25% of parosteal osteosarcomas, most commonly in recurrent tumors rather than at initial presentation. Whenever areas of dedifferentiation are diagnosed by biopsy, neoadjuvant chemotherapy may improve the clinical outcome of patients.[14] Parosteal osteosarcoma is a slowly progressive disease. Its prognosis is excellent, with 91% survival at 5 years. Pulmonary metastases tend to appear late in the course of the disease, frequently following one or more local recurrences.[15] Incompletely excised, the tumor may recur and progress to high-grade sarcoma. The presence of such areas is associated with a prognosis similar to that of conventional osteosarcoma, but better than dedifferentiation in chondrosarcoma.

In conclusion, parosteal osteosarcoma is a low-grade malignant bone tumor characterized by its insidious growth and favorable prognosis. It rarely leads to metastasis. Its treatment is mainly surgical. Our observation illustrates that this tumor is not aggressive, with no metastasis after one year of progression.

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