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# A Rare Case of Masquerading Chondrosarcoma with Pathological Fracture of Neck of Femur – A Case Report

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**Learning Point of the Article:**

Be open for surprises in cases of tumors. Always send pre-operative and post-operative biopsy to confirm your diagnosis and for further treatment.


## Abstract

**Introduction:** Chondrosarcoma is the second most common primary malignant tumor accounting for 20–27% of these. These are malignant cartilaginous tumor found most commonly in the pelvis, followed by the proximal femur. Perivascular epitheloid cell tumor (PEComa) is a rare mesenchymal neoplasm. These tumors all share a distinctive cell type, the perivascular epithelioid cell (which has no known normal tissue counter – part).

These are post commonly found in visceral sites such as gastrointestinal and uterus, in retroperitoneum and abdominal pelvic sites, rarely affecting skeletal system.

**Case Report:** A 45-ycar-old female was presented in the orthopedic outpatient department with a 2-year history of pain and swelling in her right hip with recent weight-bearing difficulties for 1 week after trivial slip and fall. X-rays revealed a lytic lesion in her right proximal femur along with neck of femur fracture. Biopsy revealed perivascular epitheloid tumor. Diagnosed with primary bone PEComa without metastasis in the right proximal femur, the patient underwent wide local excision and reconstruction using a mega-prosthesis.

Post-operative histopathological evaluation surprisingly identified a low-grade chondrosarcoma, but with clear surgical margins.

**Conclusion:** This case highlighted an unusual diagnostic discrepancy between pre-operative and post-operative evaluations. Nonetheless, the adopted strategy was effective, allowing the patient to resume daily activities. Surgeons should remain aware of the potential for primary bone tumors to masquerade as atypical tumors.


**Keywords:** Chondrosarcoma, perivascular epitheloid tumor, proximal femur, biopsy.

## Introduction

Chondrosarcoma is the second most common primary malignant tumor of bone accounting for 20–27% of these [1]. Chondrosarcoma is malignant tumor; that produces a chondrosarcoma cartilaginous matrix [2]. Classification of chondrosarcoma is based on histological subtypes, including conventional (grades 1–3), clear cell, mesenchymal, and dedifferentiated 2020 [3].

The World Health Organization classification divides chondrosarcoma into eight subtypes [4]. These are found most commonly, in the bones of the axial skeleton (pelvis, scapula, sternum, and ribs), followed by the proximal femur and proximal humerus [5]. The treatment for proximal femur tumor necessitates a two-part strategy incorporating tumor resection

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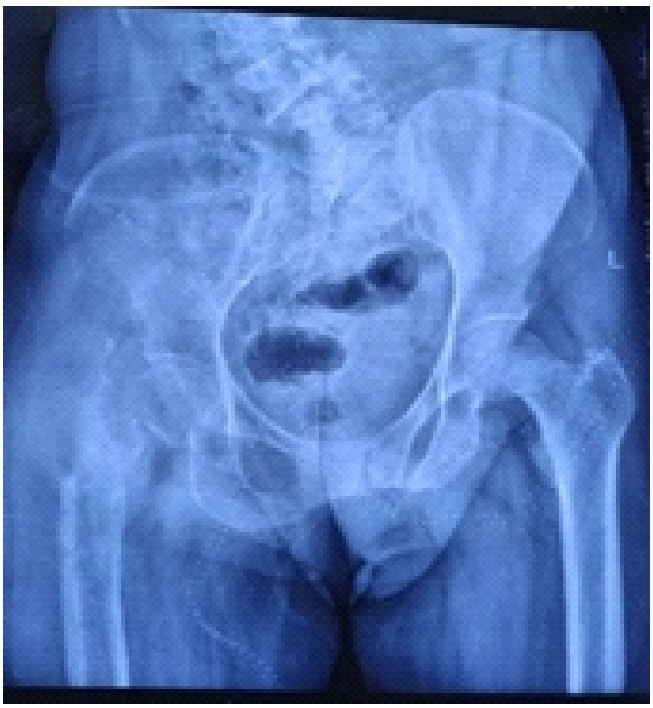

**Figure 1:** Radiograph of bilateral hip with pelvis of patient at presentation preoperatively showing a lytic lesion at right proximal femur with fracture neck of femur.

for oncological reasons and bone and soft-tissue reconstruction for mechanical purposes [6]. Perivascular epitheloid cell tumor (PEComa) is a rare mesenchymal neoplasm. These tumors all share a distinctive cell type, the perivascular epitheloid cell (which has no known normal tissue counter-part). These are most commonly found in visceral sites such as gastrointestinal and uterus, in retroperitoneum and abdominal pelvic sites [7], rarely affecting skeletal system.

## Case Report

A 45-year-old female was presented in the orthopedic outpatient department with a 2 year history of pain and swelling in her right hip with recent weight-bearing difficulties for 1 week after trivial slip and fall. The patient was unable to bear full weight on affected limb since 1 week. She faced pain at hip joint in squatting and sitting cross legged since 2 year which were gradually increasing in intensity. There was history of weight loss. There was no association with fever, chills, or history of steroid intake. Radiograph (Fig. 1) revealed a lytic lesion in her right femur, involving the entire peri-trochanteric region, whole neck, parts of femur head, and proximal shaft which was compounded by a femur neck fracture. Following initial management with traction and immobilization, further investigations, including magnetic resonance imaging (MRI) and positron emission tomography (PET) scan, were conducted. MRI (Fig. 2 and 3) revealed a T2 hyperintense, and TI hypointense, mass confined to the bone sparing surrounding soft tissues and neurovascular structures. PET scan was negative for metastasis. A core-needle biopsy indicated a perivascular epitheloid tumor. Patient was diagnosed with primary bone PEComa without metastasis in the right proximal femur. Under regional anesthesia, the patient was kept in lateral decubitus position and southern approach was used to expose lesion. The patient underwent wide local excision and reconstruction using a mega-prosthesis. Post-operative radiograph (Fig. 4) showed complete excision of lytic area replaced with mega-prosthesis appropriately. The excised tissue was postoperatively sent for biopsy to confirm the diagnosis. The biopsy result surprisingly came out as low-grade chondrosarcoma showing presence of atypical chondrocytes with hyperchromatic nuclei and calcification (Fig. 5).

## Discussion

The case report shows that sometime a common bone tumor can have atypical presentation.

Biopsy of lesion although being gold standard diagnostic tool cannot be 100% reliable due to inadequate tissue sample, wrong tissue sample or maybe due to inexperienced pathologist. PEComas are a family of mesenchymal tumors that rarely arise as a primary bone tumor [8]. Our patient bad involvement of proximal femur which is also a common location for chondrosarcoma. The age of 45 years also favors for chondrosarcoma as it is common in adults of 30–70 years of age.

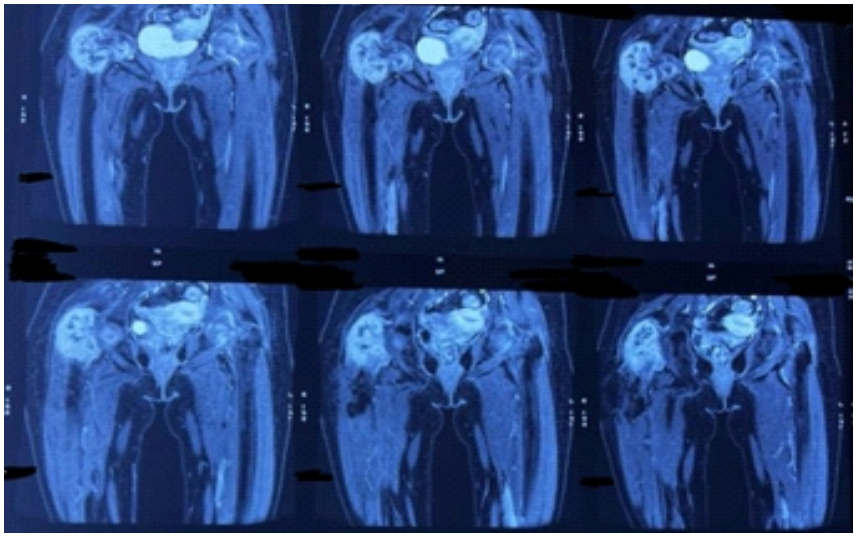

**Figure 2:** Coronal magnetic resonance imaging of bilateral hip and pelvis showing lytic lesion in proximal femur and fracture neck of femur right side.

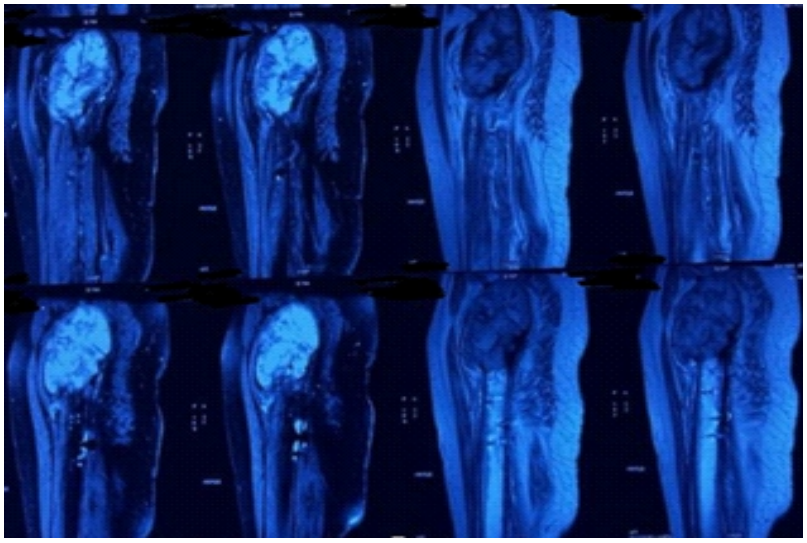

**Figure 3:** Sagittal view of magnetic resonance imaging of bilateral hip with pelvis showing extent of lytic lesion in anteroposterior plane in the right proximal femur.

Clinically, most patients present with pain, swelling, and the presence of a mass of variable duration [9]. Radiologically, chondrosarcoma shows lytic lesions (50%) intralesional calcifications: ~70% (ring and arc or popcorn calcification), endosteal scalloping affecting more than two-thirds of the cortical thickness, moth-eaten appearance, or permeative appearance in higher grade tumors, for example, myxoid, dedifferentiated, and mesenchymal chondrosarcoma. Cortical remodeling and periosteal reaction are also seen. MRI shows low-to-intermediate signal intensity lesion on T1 and T2 shows high-signal intensity lesion as was consistent with our patient [10]. Diagnosis is confirmed on biopsy. Macroscopically chondrosarcoma presents as translucent lobular, blue-gray, or white cut surface corresponding to the presence of hyaline cartilage. There may be areas containing myxoid or mucoid material and cystic changes. Microscopically, chondrosarcoma show abundant blue-gray cartilage matrix-production. Irregularly shaped lobules of cartilage varying in size and shape are present. Fibrous bands separate that these lobules or permeate bony trabeculae calcified areas suggesting the presence of a pre-existing enchondroma can often be found. The chondrocytes are atypical, with variable size and shape and contain enlarged hyperchromatic nuclei. Low-grade chondrosarcoma often closely resembles normal cartilage or the benign enchondroma, it is differentiated by presence of "chondrosarcoma permeation pattern," where the tumor infiltrates through the marrow cavity instead of being confined by the native architecture as present in benign pathology [11]. Low grade or grade 1 chondrosarcoma with no aggressive imaging features can be treated with intralesional curettage and local adjuvants [12]. However, when aggressive biologic behavior is evident on imaging, wide resection following surgical principles of malignant bone tumors seems more appropriate. Our patient underwent wide resection and reconstruction with mega-prosthesis as the pre-operative biopsy diagnosis was PEComa which needs wide surgical excision [13]. However, despite the inconsistency in diagnosis, the patient had optimum final outcome.

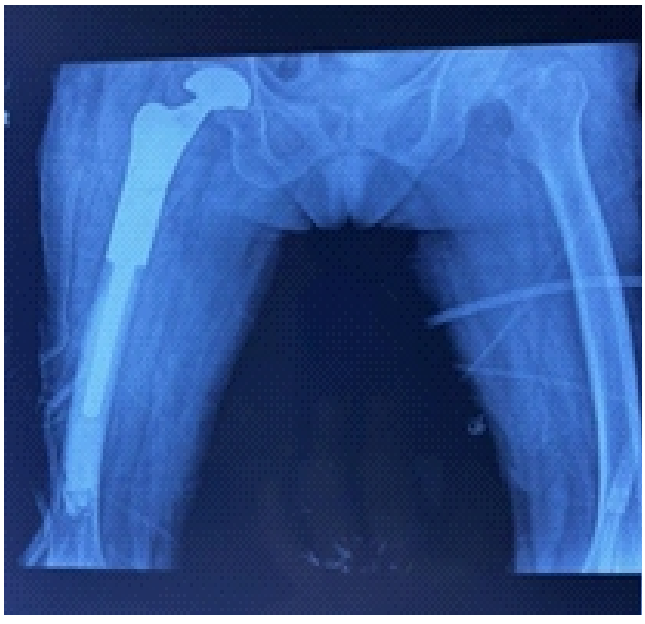

**Figure 4:** Post-operative radiograph of patient showing removal of lytic lesion and replacing proximal femur with mega prosthesis.

## Conclusion

This case highlighted an unusual diagnostic discrepancy between pre-operative and post-operative evaluations. Nonetheless, the adopted strategy was effective, allowing the patient to resume daily activities. Surgeon should always send

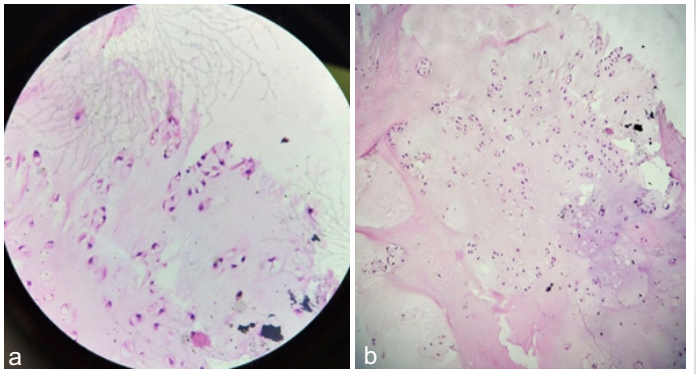


**Figure 5:** (a and b) Post-operative histopathological report showing grade 1 chondrosarcoma.

post-operative tumor sample for confirmation of diagnosis and if there is discrepancy in diagnosis do the needful for the well-being of the patient.

**Clinical Message**

Surgeons should remain aware of the potential for primary bone tumors to masquerade as atypical tumors.

**Declaration of patient consent:** The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given the consent for his/ her images and other clinical information to be reported in the journal. The patient understands that his/ her names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Conflict of interest:** Nil **Source of support:** None

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