Case report

Clear cell chondrosarcoma proximal femur with secondary aneurysmal component – A rarity

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1. Introduction

Clear cell chondrosarcoma is a rare benign tumor constituting to about 2–4% of all chondrosarcoma. It was first described by Unni et al. in 1976¹. Common sites of occurrence are epiphyseal metaphyseal regions of femur, proximal humerus and proximal tibia. This tumor clinico radiologically is frequently misdiagnosed as benign lesion such as giant cell tumor, aneurysmal bone cyst, chondroblastoma & osteoblastoma. Conservative surgeries like curettage will lead to local recurrences, excision of the lesion & reconstruction is the treatment of choice. Despite low malignant potential, recurrence & metastasis have been reported.

2. Case report

A woman aged 55years attended orthopedic outpatient department with complaints of pain in right hip since 2 years. To start with symptoms were of mild to moderate grade. For past three months symptoms got aggravated hindering daily activities. There was no history of trauma, fever or massages. No history of steroid intake or contact history of TB. Examination revealed mild fullness in right groin with a tender hip anteriorly. Movements were grossly restricted concentrically and were painful. No other joint involvement was appreciated. With clinical suspicion of avascular necrosis with secondary osteoarthritis, patient was further evaluated. Routine investigations were within normal limits. She was diagnosed as diabetic on admission. Alkaline

phosphatase & Calcium levels were within normal range. Patient had normal lipidemic levels and negative sickling test.

Abdominal Sonography revealed no organomegaly.

Radiographs revealed multiple cystic areas with mild sclerosis in right femur head with expansile lytic lesion in neck extending into intertrochanteric region obliterating head & neck offset. No matrix calcification (Fig. 1).

BONE SCAN revealed increased uptake in right head & neck femur with no evidence of increased uptake in other parts of skeleton.

CT revealed mildly expansile lytic lesion in head & neck femur with clear borders. No matrix calcification (Fig. 2).

MRI revealed low signal intensity in T1 Weighted images, moderately bright signals in T2 Weighted images. No soft tissue involvement (Fig. 3).

As none of the radiological investigations were confirmatory, patient was subjected to FNAC. FNAC under fluoroscopic guidance revealed features suggestive of Aneurysmal Bone Cyst.

The lesion being juxtaarticular with no cortical breach or soft tissue involvement patient was planned for excision and hemiarthroplasty. Intra operatively there was no evidence of pathological fracture or soft tissue extension, head & neck ballooning was seen. Intact head was dislocated and excision of head and neck was done 1 cm proximal to the lesion. Proximal femur was replaced with HA coated modular bipolar stem. Excised specimen was sent for Histopathological examination. Postoperative period was uneventful. Sutures were removed on 10th postoperative day and gradual weight bearing was started with walker. Later walking without aid was permitted by 6th post-operative week. At recent follow up of 1 year, patient was walking without support and pain. Gait patterns were normal, no limb length discrepancy and hip range of movements were near normal.

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Fig. 1. X ray shows very well defined lytic lesion seen extending from neck of right femur to right intertrochanteric region inferiorly. No obvious calcification. Sharp zone of transition noted with mild sclerotic border. Sub articular cystic changes in right femur head which appears mildly sclerotic.

Fig. 2. CT scan shows mildly expansile lytic lesion without obvious matrix calcification with thinning of cortex. Lesion is involving epiphysis & extending into intertrochanteric region with sub articular cortical breach at few levels.

Cytology of sections showed lobules of low grade mesenchymal tumor cells composed of large, round cells with clear cytoplasm & centrally placed nuclei. Tumor lobules exhibit few small cystic spaces surrounded by multinucleate giant cells. In centre of lobules, spicules of calcification present. There are areas

resembling ABC. Histopathological report was opined as clear cell chondrosarcoma with Aneurysmal Bone Cyst like component (Fig. 4).

Fig. 3. MRI shows on fat saturated proton density significantly hyper intense lesion. No perilesional edema. No soft tissue extension. Radiologically this has been diagnosed as benign chondrogenic lesion/giant cell tumor.

Fig. 4. microphotograph showing sheets of round to ovoid tumor cells with abundant clear cytoplasm and minimally pleomorphic round hyper chromatic nuclei along with woven bone and osteoclastic type of giant cells (H & E, ×200).

3. Discussion

Clear cell chondrosarcoma is a rare low grade malignant tumor that was first described by Unni et al. in 1976¹. It has an incidence of 2–4%² more so is the rarity when associated with ABC like components.³ It usually occurs in 3rd to 5th decade of life.⁴ Commonly seen in third to fifth decade of life. Males are affected twice as common as females. Most of these tumors occur in epiphyseo metaphyseal region of long bones such as femur, tibia and proximal humerus. Other rare sites include skull, spine, pelvis, phalanges, and larynx.⁵

Radiologically these tumors appear as expansile, radiolucent lesions in epiphyseo metaphyseal region with intact cortices.⁶ No periosteal new bone formation or matrix calcification is seen. Cartilage tumors generally exhibit matrix calcification in the form of rings or popcorn shape, clear cell chondrosarcoma is an exception. Diaphysis is rarely involved⁷. These nonspecific features commonly lead to misdiagnosis of benign lesions such as Aneurysmal Bone Cyst, chondroblastoma, Giant Cell Tumor, osteoblastoma.

Even though clear cell chondrosarcoma resembles chondroblastoma microscopical presence of clear cells make them unique. Most of the clear cells on electron microscopy show irregular shaped nuclei with indentations. Large dilated endoplasmic reticulum cisternae, bundles of actin like filaments & a few glycogen particles.⁶ Confusion with osteoblastic neoplasms may be clarified on electron microscopy by showing that cells lining the bony spicules are not part of the neoplastic process but rather are the result of stromal reaction.² Presence of clear cells is the primary differentiating feature between giant cell tumor/aneurysmal bone cyst and clear cell chondrosarcoma.² Rarely aneurysmal bone cyst like features radiologically and microscopically may co exist with clear cell chondrosarcoma as seen in our case, these features depend on the phase of presentation.³

Serum calcium, phosphorus will generally be within normal range. Alkaline phosphatase may be within normal range or elevated. Elevated levels of Alkaline phosphatase on follow up is a warning sign signifying recurrences or metastasis.⁸ Recurrences as late as 29 years post excision of primary lesion has been reported.⁵ Our case was followed up to 1 year and we observed no local recurrence or distant metastasis

Histologically clear cell chondrosarcoma exhibit lobules of cells with clear cytoplasm. Well defined cytoplasmic borders and central round nuclei with vesicular chromatin. Benign osteoclast like giant cells may be seen at periphery. Fine lines of calcification, osteoid or bone formation may be found scattered among tumor cells. Chondroid matrix may also be present.^{3,9}

Despite the presence of a low malignant potential these tumors are not to be treated by curettage and grafting as it has a high recurrence rate. Wide excision and reconstruction is the treatment of choice. Other options include en bloc resection including a margin of normal bone.

4. Conclusion

In conclusion clear cell chondrosarcoma is a rare low grade malignant bone tumor occurring in epiphyseo metaphyseal region. Radiological features mimic a giant cell tumor, aneurysmal bone cyst, chondroblastoma & osteoblastoma. To prevent confusion in diagnosis cytopathology should always be considered in every case. Though low grade malignant, these tumors are to be treated by radical excision and reconstruction to prevent recurrences. Patients are to be regularly followed periodically as recurrences and metastasis may occur at a later date.

Source of support

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Conflict of interest

None.

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