CASE REPORT

MULTICENTRIC SYNCHRONUS GAIANT CELL TUMOR: A CASE REPORT

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INTRODUCTION: Giant cell tumors are relatively common primary bone tumors, and represent 4-5% of all primary bone neoplasms. They are locally aggressive lesions and tend to metastasize. They commonly present as pain and swelling in the metaphyseal or subarticular region in 3rd and 4th decades of life with female predominance. The distal femur, proximal tibia (Around the knee joint) and distal radius are the most common sites of involvement and account for 50% of cases.

CASE HISTORY: A 34 years old female patient presented with slowly growing painful swelling just below the right knee of more than 6 months duration. There was no history of trauma, fever, other constitutional symptoms and diabetes. On physical examination, an oval firm swelling of 8x5x6.5 cm size with stretched skin over the swelling without ulceration or inflammatory signs was present on the anterior lateral portion of upper leg just below the knee joint. Minimal restriction of extension and rotational movements were noticed. There was no lymphadenopathy and no other bony or soft tissue swelling were identifiable in rest of the body.

Routine blood investigations such as Hemogram, ESR, CRP, ASO titres, uric acid Serum calcium, phosphorus and alkaline phosphatase levels were within normal limits. No focal lesions were found on chest radiograph.

Plain Radiograph of knee revealed subarticular expansile osteolytic lesion in tibia and fibular head with narrow zone of transition, destroyed trabeculae and without matrix mineralization. Cortical breach and periosteal reactions were not observed. (Fig. 1).

Based on the radiographic features Giant cell tumor and Brown tumor of hyperparathyroidism were thought as probable diagnosis. Brown tumor was excluded after finding normal parathyroid glands on ultrasonogram of neck and normal levels of serum parathormone and serum calcium levels.

MDCT and MRI were done to evaluate the cortical breach, articular and soft tissue involvement before definitive treatment planning. In addition to radiographic findings, the lesions in tibia and fibula are not separable from each other on MDCT and MRI (Fig. 2, 3). Cortical breach and soft tissue extension were observed at several places on MDCT and MRI (Fig. 2, 3). Fortunately there was no intra articular extension into the knee joint.

Excision of the fibular head with curettage and bone grafting of the tibial lesion were done and specimens were sent for histopathological examination separately. HPE sections revealed linear fragments of tumor tissue showing mostly stromal component with spindle cells arranged in loose fascicles having uniform nuclei. Few scattered multinucleated osteoclast like giant cells were present. In addition few islands of irregular bony trabeculae with no osteoblastic rimming were also seen. No significant mitotic figures were present and all the features were consistent with giant cell tumor for both fibular head and tibial lesion. So finally a diagnosis of synchronous multicentric giant cell tumor involving tibia and fibula was made. Patient was on regular follow up since 1 year and till now no local recurrence or metachronous appearance of new lesions was observed.

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DISCUSSION: Gaint cell tumor are locally invasive tumors.⁽¹⁾ They form 4 % all primary bone tumors and 10 % of all benign tumors with female to male ratio of 3: 2. Most of them are benign tumors with locally invasive nature. Malignant Gaint cell tumors do novo are rare and the so called malignant Gaint cell tumors are really osteoclast rich osteosarcoma, fibrosarcoma or represent post radiation sarcomas. In 10% of patients pathological fracture may occur.⁽²⁾ Gaint cell tumors can also manifest as regional and systemic tumor implantation, and malignant transformation with metastases to lungs in 2% to 5% cases.⁽¹⁾ The rate of recurrence post treatment is 5 % to 10 % with modern therapeutic methods such as wide excision, cryosurgery, bone grafting and radiotherapy.⁽¹⁾

Whereas MCGCT is extremely rare and account for less than 1 % of Gaint cell tumor.⁽³⁾ Involvement of metaphyseal region, hand bones and pathological fractures are common in MCGCT.⁽¹⁾ In the literature isolated single case reports of MCGCT were there but Hoch B et al⁽³⁾ reported single large series of 30 cases of Multicentric gaint cell tumors with clinico pathological analysis. The mechanism of multicentricity was postulated by Dhillon et al⁽⁴⁾ as contiguous spread, iatrogenic tumor cell seeding, benign metastasis, malignant transformation and de novo formation either synchronously or metachronously.

In our case both tibia and fibula are involved synchronously, may indicate local extension of tibial lesion to fibula or vice versa. There was no regional or systemic involvement in our case.

Hoch B et al reported majority of MCGCT appearance in younger patients with average age of 21 years and female/ male ratio of 2:1⁽³⁾ with an average of three tumors per patient and most of their location in long bones around knee, proximal humerus and distal radius. In the same series six patients had synchronous and rest of them had metachronous lesions and most of them appeared in subarticular location, whereas in immature skeleton the lesions were noticed in metaphyseal region. Most tumors were treated with curettage (64%) or resection (22%) in their series with metastatic disease in three and malignant transformation in two. Unlike Hoch B series our case is a synchronous gaint cell tumor of tibia and fibula appeared in slightly older age female of 34 years. Similar to Hoch B series our patient also underwent resection of fibular lesion and curettage of tibial lesion with bone grafting and no recurrences was observed till 1 year post surgery. Fibula involvement is rare in MCGCT and only 4 tumors involved fibula out of 94 tumour in the Hoch B series. But in our case fibula involvement was present.

Dhillon et al reported involvement of short bones of hands and feet and metadiaphyseal region of long bones as common site for MCGCT⁽⁴⁾ but in our case subarticular region of tibia and fibular head are involved.

Hoch B et al reported two or three lesions per patient in their series of 30 cases⁽³⁾ but Park IH et al reported ten lesions in a single patient.⁽⁵⁾ In our case only two lesions were detected. Aneurysmal bone cyst, brown tumors, chondroblastoma and intra articular ganglion are considered under differential diagnosis and they can easily be differentiated by clinical and radiological features.⁽⁶⁾ Aneurysmal bone cysts are located centrally in metadiaphyseal region of long bones before epiphyseal fusion. Brown tumors are also in metadiaphyseal in location and excluded by clinical features, hyper calcaemia and parathormone estimation. Chondroblastomas are in epiphyseal location in immature skeleton with internal calcifications. Intra articular ganglion cysts commonly occur in medial malleolus and in carpal bones.

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For treatment planning magnetic resonance imaging (MRI) is the modality of choice to assess the actual tumor extent in the bone and in the soft tissues and for assessment of articular surface Involvement.⁽⁶⁾ CT is a reasonable alternative.⁽⁵⁾

The radiographic features, cytomorphology and histopathological characters of MCGCT do not differ from that of solitary tumors. However fibroblastic and fibrohistocytic areas were present in some cases of metaphyseal lesions of Hoch B et al series.⁽³⁾ Treatment of MCGCT may differ significantly from that of solitary lesions. Each lesion must be evaluated and treated individually. Some lesions may show indolent course with no change over long periods of time; in such cases patient can be followed up radiologically without treatment, till functional disability or aggressive features become apparent. The response to treatment and recurrence rates are similar in both MCGCT and isolated cases.⁽⁷⁾ Patients with MCGCT require long term follow up due to metachronous lesion development. Bone scintigraphy is very useful in the follow up of individuals with MCGCT in detecting new lesions and excluding recurrent disease.

CONCLUSION: MCGCT are rare and can be synchronous or metachronous and occur at earlier age than in solitary Giant cell tumor and involvement of a fibula is again rare among all MCGCT sites. Our case is a synchronous MCGCT in a 34 years old female, located in the commoner site for giant cell tumors around the knee joint. Though fibula involvement is rare in MCGCT both fibula and tibia are involved in our case.

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Fig. 1: X –ray Right knee AP and Lateral views showing Expansile lytic subarticular lesions involving proximal Tibia and Fibular head

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Fig. 2: Axial CT images showing expansile lytic lesions in Proximal tibia and Fibular head with multiple cortical breaks

Fig. 3: Coronal and Saggital MRI images demonstrating Expansile lesions in Proximal tibia and Fibular head with Fluid - fluid levels and cortical breach

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