Case Report

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# Dedifferentiated Low Grade Chondrosarcoma Of Calcaneum A Rare Tumor: A Case Report

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## Abstract

**INTRODUCTION**

Tumors of calcaneum are very rare. Due to lack of awareness about the calcaneum tumors among the general physicians, there is always either delay in diagnosis or misdiagnosis and therefore there is dilemma in offering correct treatment. Here we are presenting a rare case of dedifferentiated mild grade chondrosarcoma of right calcaneum, which was initially diagnosed on FNAC as Chondromyxoid Fibroma.

**CASE REPORT**

A 71-year-old man referred to OPD by local practitioner, for right heel pain on and off and swelling for two year which was not responding to the treatment. Swelling around the heel was gradually progressing. X ray of right foot and ankle showing soap bubble appearance in the calcaneum and cortex was intact. Patient underwent FNAC and the report was suggestive of chondromyxoid fibroma a rare benign tumor. Hence patient underwent excision, curettage and bone grafting procedure. The material sent for re- confirmation and the report was confirmed as dedifferentiated type of low-grade chondrosarcoma, a low-profile malignant tumor. Below knee amputation was carried out. Patient now, six-month post-surgery and walking on 'Jaipur foot' type of prosthesis with no relapse till date.

**CONCLUSION**

Tumor of calcaneum is very rare, hence detail clinico-pathological-radiological evaluation is very much essential to avoid misdiagnosis and delay in treatment. Chronic heel pain and swelling around heel are misdiagnosed as chronic benign conditions like ankle ligament sprain or retrocalcaneal bursitis, plantar fasciitis, infection or old malunited fracture calcaneum. Any tumor of calcaneum which is rare should be reported and multidisciplinary evaluation to be carried out along with limited open biopsy (Gold standard), to confirm the histological diagnosis.

**Keywords:- Heel Pain, Chondro-Myxoid Fibroma, Chondrosarcoma, Amputation.**



## INTRODUCTION

Calcaneum is largest tarsal bone in the foot. It is developing during 4th-7th week of gestational life.[1] It is the second commonest site in the foot after metatarsals for tumor.[2] Malignancy of foot reported as less than 3%. Calcaneum tumor reported as 31% benign and 35% malignant. Dull aching pain and gradual onset of swelling are the most common types of clinical presentation in absence of trauma. Rarity, lack of knowledge, delay in diagnosis, chances of misdiagnosis and inadequate treatment reported in literature.[3]

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Most of the clinicians and pathologist are unaware of existence of calcaneum tumor and the chronic heel pain and swelling misdiagnosed as plantar fasciitis, ankle sprain, retrocalcaneal bursitis, Os trigonum, pump bump and malunited fractures and many more. There are both, common and rare types of tumors of calcaneum reported in literature. Here we are reporting one of such a case with heel pain and swelling who was treated initially by local practitioner with analgesics and later referred to us for further management. After studying the case, we first thought of benign tumor and carried out FNAC which shows rare benign tumor and treated with curettage and bone grafting. The curated material reported dedifferentiated low grade rare malignant tumor, with MRI showing no distant spread, hence below knee amputation was later carried out. In a rare site like calcaneum tumor, local minimal open biopsy to be considered because of higher accuracy rate (98%) compare to FNAC (96%) to avoid mis-diagnosis and delay in treatment.

# CASE REPORT

71-year-old elderly person referred to us for evaluation of chronic pain and swelling at right heel for 2 years with no history of trauma, morning stiffness, loss of appetite or weight loss or evening rise of temperature. No history of sleep disturbance at night due to pain. No significant personal or family history. The pain was insidious in onset, dull aching type, localized to right heel aggravated by walking and relieved on rest. Swelling around ankle joint was diffuse type gradually progressing, firm in consistency with no crepitus. Diffuse tenderness was there on palpation over the calcaneum. skin over the calcaneum was intact with no evidence of scar, sinuses or any discharge. No evidence of dilated veins over the swelling and no evidence of any lymphadenopathy anywhere in the body. There was no involvement of other major or minor joint. There was no evidence of any systemic involvement. Chest and per abdominal examination were normal. Radiological evaluation shows soap bubble appearance type of lytic lesion at the calcaneum with cortex intact. Other foot bones were normal. Blood investigations were normal and tumor markers were negative. X ray chest and ultrasonography of abdomen was normal.

After complete clinical evaluation, FNAC of the lesion over the calcaneum was carried out which showed chondromyxoid fibroma, a rare tumor of calcaneum. Considering all clinic-radio-pathological evidence excision, curettage and bone grafting treatment option was chosen and the procedure was carried out. The curated material was again sent for the histopathological evaluation and then it was reported as low grade de-differentiated chondrosarcoma, type of malignant tumor. MRI of tibia, foot & HRCT CHEST was carried out. As there was no distant spread in tibia proximally, below knee amputation was carried out. Patient now is 6-month post operative, wound healed primarily and patient is wearing Jaipur foot type of prosthesis and rehabilitated full weight bearing along with cane support. There is no relapse or secondary growth of tumor till date.

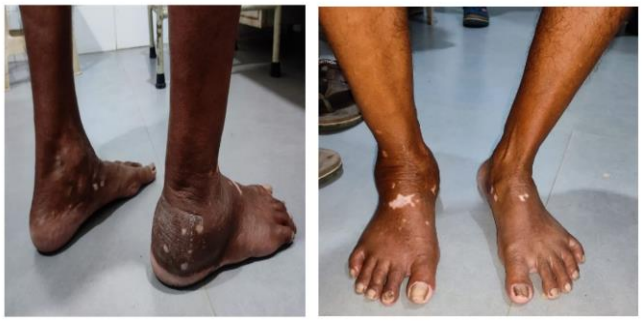

**Figure 1:- Clinical Image showing distinct swelling around right ankle.**

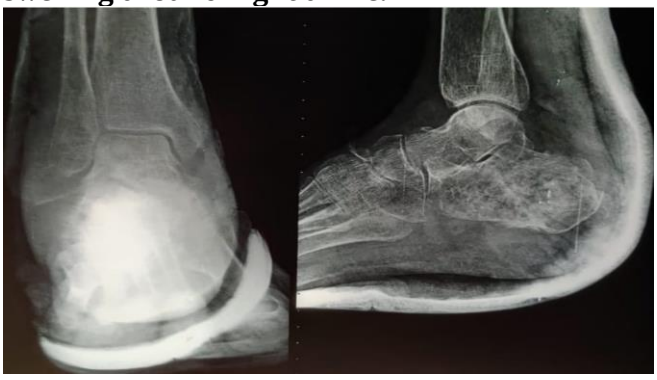

**Figure 2:- X-Ray of calcaneum showing characteristic soap bubble appearance**

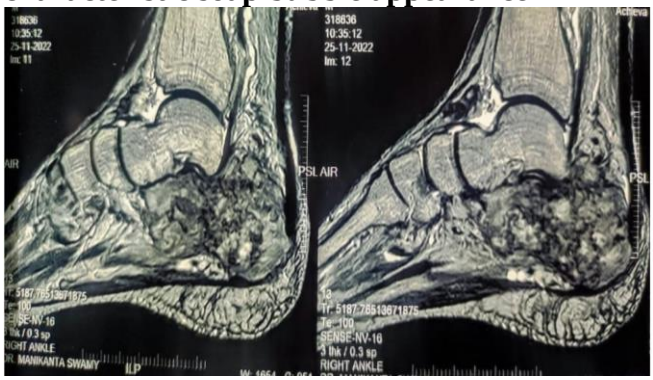


**Figure 3:- MRI foot showing absence of tibial involvement.**

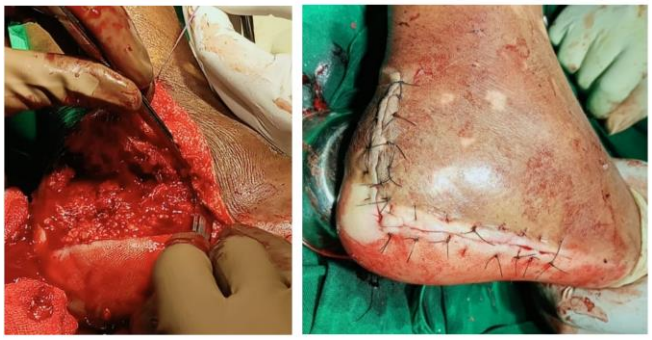

**Figure 4:- Bone grafting and closure of the wound**

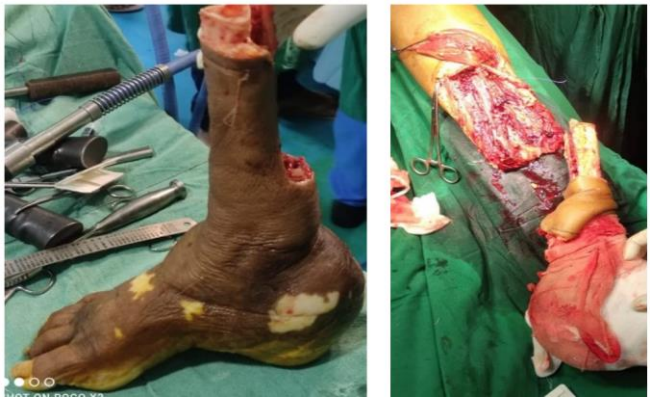

**Figure 5: Below knee amputation procedure**

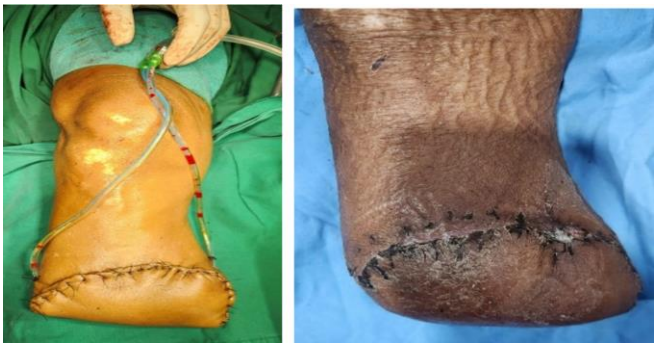

**Figure 6: Amputated stump and wound healing**

**Figure 7:- Rehabilitation of the patient by using Jaipur foot type of prosthesis.**

# DISCUSSION

Tumors of foot are very rare. Tumors of calcaneum are mistreated by clinicians, as chronic medical conditions due to lack of awareness and experience. Any heel pain or swelling in absence of significant trauma, which is not responding to treatment, should be evaluated properly. A multidisciplinary approach involving clinico-radio-pathological evaluation should be considered. Therefore, we are discussing here, some of chronic medical conditions related to calcaneum and ankle, benign and malignant tumors of calcaneum and their characteristic features to enhance our knowledge further. It will help us diagnosing and evaluation of these conditions correctly.[4]

**Benign chronic medical condition[5]**

**Plantar fasciitis**—Anterio medial plantar aspect of heel pain classically seen in athlete and patient with walking or standing for long hours. Hypothyroidism, rheumatoid arthritis and Diabetes mellitus are associated clinical conditions. Radiologically calcaneum spur seen most of the time. This condition is treated conservatively with heel cushion, sports shoes, physiotherapy in the form of ultrasonic waves and steroid injections or platelet rich plasma injections.[6]

**Os trigonum**- It is common in ballet dancers and sports person with using foot in hyperflexion position. There is small bony outgrowth on posterior aspect of talus which causes impingement on plantarflexion of ankle. This condition is to be treated conservatively with rest, physiotherapy and lifestyle modification.[7]

**Ankle ligament sprain**-Patient has history of twisting of ankle joint and pain on the side of ankle joint, depending upon the side involvement. Anterior Tibiotalar ligament sprain is most common. Treatment is conservative management with rehabilitation. If not treated properly then result in chronic pain, swelling, instability and stiffness**.**

**Heglund's deformity (Pump bump)**-This is a chronic condition where there is bony bump present on posterior superior aspect of calcaneum This is due to irritation of the shoes or it may be because of genetical preponderance. Initial phase conservative management, if no relief then surgical excision of pump bump needed.[8]

**Retrocalcaneal bursitis**-This is a condition where there is inflammation of the bursa lies superior aspect of calcaneum. There is pain, irritation and swelling on both side of Tendo achillis tendon. Initially treated with analgesics, physio, rest, shoes

modification. If no relief, then surgically removal of the bursa is advised. If not treated properly then it will get converted into the chronic condition with pain and swelling around ankle.[9]

**Fracture of calcaneum -** Malunited fractures (extra articular tounge type or intra articular) of longer duration, present with chronic pain and swelling at heel area. Pain more in walking and relieved on rest. Stiffness is present at subtalar joint and ankle joint. Local pain management, exercise, shoe modification, intra articular steroids or platelet rich plasma (PRP) injections are the treatment options. In case of severe arthritis, fusion of the subtalar joint advised.

**Benign tumors of calcaneum**

**Simple bony cyst**—Less than 3% of all osseous tumors.14% morbidity reported in literature. Most common site is Ward 's triangle which is located anterolateral to posterior articular facet where the stress is minimum. Macroscopically fluid filled cavity separated by fibrous membrane. Microscopically cholesterol cliff is present which separates it from other bone cyst in long bones where it is absent. Treatment is by calcium sulphate cement, ceramics, Tri calcium phosphate, steroid injection or bone grafting. Post op no recurrences reported in literature.[10]

**Aneurysmal bony cyst-** less than 1% incidence among all osseous tumors.30-35% associated with other bone tumors like giant cell tumor, osteoblastoma, chondroblastoma. Chronic heel pain is the main clinical feature. On x ray, lytic expansile lesion seen. On MRI multiple free fluid level seen. Histopathology shows histiocytes, osteoclasts, fibroblasts. Treatment option is curettage and bone grafting. Cytometry, carbolic acid, phenol therapy, methylmethaacrylate bone cement (MMC) and irradiation are the other treatment options available.[11]

**Osteoid Osteoma**-More common in younger population 7-25 age groups. More common in males reported in literature. Clinically nocturnal pain and swelling and physical examination is generally nonspecific. X ray shows sclerotic lesion. CT scan shows central nidus with surrounding sclerosis. MRI is reserved for more symptomatic patients. Histopathology shows that the nidus consists of osteoblast, osteoclast surrounded by giant cells with fibrous and vascular tissue. Treatment first with NSAID if no relief then curettage and bone grafting. CT guided resection, LASER photo coagulation are other noninvasive methods of treatment described in literature. [12]

**Osteochondroma**-10% of all osteochondromas reported in hands and foot. If age less than 20 at the time of presentation, malignant transformation in future possible as reported in literature. Clinically pain, swelling and stiffness present in foot. Stiff, nodular, painful mass present then malignant changes can be possible. X ray shows sessile or pedunculated mass. Hyaline cartilage cap not seen on x ray. On MRI, hyaline cartilage cap can be seen. The cartilage cap thickness more than 1.5 cm suggestive of malignancy. Histopathology shows spongy mass covered with hyaline cartilage cap. In symptomatic patient excision of osteochondroma to avoid malignant transformation is suggested in literature. [13]

**Chondroblastoma-**4.5 % of all tumor types. 2.8% of all calcaneum tumor type. M: F sex ratio is 5:1. Clinically pain and soft tissue swelling. x ray shows osteolytic lesion MRI shows multiple free fluid levels Histopathology shows polygonal chondroblast, multinucleated giant cells and calcification. Surgery is simple curettage and bone grafting. [14]

**Intraosseous lipoma**-incidence 0.1% over all. 8% percent located in calcaneum. Clinically pain on walking and relieved by rest NSAID Lab reports normal X rays shows well circumcised lytic lesion with central ring like calcification located in the neck region of Ward's triangle. Gross appearance shows yellow friable tissue and microscopically fat cells surrounded by necrosis and calcification and hemorrhage. Treatment option in symptomatic patient is curettage and bone grafting and long duration below knee plaster cast and non-weight bearing for six weeks. No recurrence is reported in literature after curettage.[15]

**Giant cell tumor**-1.2% of all tumors of calcaneum. High rate of recurrence and potentially aggressive tumor 30-40 years is a common age group of presentation. Heel pain and swelling are common complaints. Radiologically, eccentrically located, expansile lesion, along with non-sclerotic margins. Histopathology shows multinucleated giants' cells and spindle cells. Treatment option is curettage with phenol, carbolic acid and bone grafting. Complete calcaneoctomy and defect filled with sural Flap is an alternative to amputation, reported in literature.[16]

**Chondromyxoid fibroma**- <7% incidence. Slow growing tumor. Most common in proximal tibia 53%, distal femur 27% and fibula 20%. **20-25%** cases of chondromyxoid fibroma, misdiagnosed in literature. Clinically chronic heel pain of longer duration and limping is present along with slow growing tumor mass. Laboratory investigations normal. X ray shows well defined geographic lesion with thin layer of sclerotic margin with well-defined septation and lobulation. CT scan shows expanded and thin cortex. RI shows T1 low intensity image and T2 high intensity image because of chondroid and myxoid composition. Gross appearance shows grey on white mass(multilobulated) with fibro gelatinous and cartilage mixed material. Histopathology, shows intra cellular margin of combination of chondroid and myxoid matrix with spindle shaped cells, stellate cells and varying number of multinucleated giant cells. Presence of large pleomorphic cells confused for chondrosarcoma. In literature, Jamshidi et al presented largest series of chondromyxoid fibroma of calcaneum of five patients. Treatment option is simple bone curettage and bone grafting and it is an effective treatment reported in literature and amputation reserve for locally aggressive tumor.[17]

**Malignant tumors of calcaneum**

**Osteosarcoma-**1% of all osteosarcoma. 10-30 years is the common age group. In case of small cell osteosarcoma there is increased CRP and ESR.X ray shows classical sun burst appearance, periosteal reaction, Codman's triangle, sclerotic reaction, cortical destruction and soft tissue expansion. Gross appearance white yellow heterogenous smooth firm mass located in the body of calcaneum. Histopathology shows spindle cells and atypical cells with lots of osteoid deposits. Treatment depends upon Ennking classification stage at the time of presentation. Chemotherapy, below knee amputation and prosthetic replacements are treatment of choice.[18]

**Primary osseous lymphoma**-Approximately 1.2% reported in literature. Clinical presentation is nonspecific Heel pain, swelling with lack of constitutional symptoms with chances of pathological fracture. Laboratory tests normal. X ray shows moth eaten appearance, patchy appearance, periosteal reaction, pathological fractures and soft tissue infiltration. Histopathology shows large number of lymphoid clusters, hyperchromatic large nuclei, abundant cytoplasm and prominent nuclei seen**.** 'Suet' like tissue in case of T cell lymphoma and CD 20 and L26(a pan Beta cell marker) present in case of B cell lymphoma. Chemotherapy and radiotherapy are the treatment of choice.[19]

**Chondrosarcoma**- less than 3% incidence in the foot.25% in calcaneum. Insidious onset, gradually increasing, diffuse swelling, increased local temperature X ray shows bony expansion, cortical destruction, endosteal erosion. Periosteal reaction present. Pathological fractures present. CT scan shows cortical destruction. MRI shows T1 low intensity and T2 high intensity images. Lobulated growth pattern. Macroscopically shows yellow color tumor mass and microscopically it shows pleomorphic chondrocytes, hyper cellularity, cytologic atypia. Cytochemistry shows tumor cells positive for S-100 protein. Treatment option wide excision or amputation. This tumor is radio-chemo resistant. Recurrence and metastasis are common at proximal end of long bones and lungs. Prognosis is guarded.[20]

**Ewing sarcoma**- More than 50% of all cases of Ewing sarcoma reported in foot. Clinically, Local pain swelling, erythema, fever weight loss, shiny skin with dilated veins. Increased ESR, acid phosphatase. X ray show moth eaten appearance, onion peel appearance. MRI T1 image shows low intensity and T2 image shows increased intensity. Bone scan shows increased uptake. CT chest shows both micro and macro metastasis. Histology shows malignant small round cell tumor CD 99 and periodic acid Schiff test positive on histochemical analysis Treatment is amputation, chemo and radio therapy. Prognosis is worst. [21]

Other very rare malignant tumors of calcaneum reported in literature are malignant fibrous histiocytoma, multiple myeloma, haemangio-endothelioma, fibrosarcoma, malignant peripheral nerve sheath tumor, primary epithelioid angiosarcoma, primitive neuro ectodermal tumor and adamantinoma.

Dealing with chronic pain and swelling at heel or any suspicions for tumor, prompt evaluation should be carried out and treatment should be given. FNAC has over all accuracy rate 96.3% and open biopsy accuracy rate is 98% as reported in literature.

# CONCLUSION

Calcaneum tumor are very rare. Patient with chronic heel pain and swelling in absence of trauma, should be evaluated in detail. Due to poor awareness about calcaneum tumor, misdiagnosis and delay in treatment is common among the clinicians and pathologists. A Clinico –Patho-Radiological multidisciplinary evolutionary, approach must be considered for evaluation of such cases. For tumor of calcaneum, because of rare site and because of higher accuracy rate, limited open biopsy method (gold standard) to be considered to avoid misdiagnosis. To improve awareness among the clinicians-radiologists and pathologists, frequent case reporting, periodic discussion in clinical meetings and tumor related clinical research of calcaneum should be carried out in future.

## Conflict of interest

None


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None


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