Extra articular Extra-synovial Solitary Osteochondromatosis of the Ankle

S Madi\textsuperscript{1}, S Vijayan\textsuperscript{2}, MA Naik\textsuperscript{3}, SK Rao\textsuperscript{4}

ABSTRACT

Synovial chondromatosis or osteochondromatosis is a benign neoplastic condition arising from synovial tissue of joints, tendon sheath and bursa. Commonly involved joints are the knee, hip, shoulder, elbow and ankle. According to authors’ knowledge, only four cases have been reported in the English literature describing the extra articular synovial chondromatosis around the ankle joint. The peculiarity of the index case lies in its subtle clinical and radiological presentations which can create a diagnostic dilemma.

Keywords: Ankle, extra-articular; solitary, synovium, osteochondromatosis

From: Department of Orthopaedics, Kasturba Medical College, Manipal, Manipal University, Karnataka, India 576104.

Correspondence: Dr S Vijayan, Department of Orthopaedics, Kasturba Medical College, Manipal, Manipal University, Karnataka, India 576104. E-mail: sandeep.vijayan18@gmail.com

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INTRODUCTION

Synovial chondromatosis is a condition characterized by the formation of multiple osteochondral loose bodies in the joints. Clinically, these loose bodies can lead to locking of the joints or restrict the range of motion. Radiological picture is not only characteristic but almost diagnostic. There have been only four case reports till date describing the extra articular synovial chondromatosis out of which two were extra articular and two had both extra as well as intra articular involvement (TABLE). Despite rarity of this condition in the ankle region, in all these cases the clinical and radiological signs were certain give away towards the diagnosis thus resulting in prompt management. Here, we describe a case of extra articular extra synovial solitary chondromatosis of the ankle, the diagnosis of which was delayed due to subtle presentation.

CASE REPORT

A 49 year old female without pre-existing co-morbidities presented to our foot and ankle clinic with complains of dull aching pain in her right ankle of more than three years duration. For the past one year, she also noticed a soft swelling over the lateral aspect of the ankle. She consulted a general practitioner who treated her with pain killers and herbal medicines diagnosing it as degenerative osteoarthritis of the ankle joint. The pain subsided with medications and rest but aggravated on prolonged standing and walking on uneven ground. There was no history of trauma, constitutional symptomsor involvement of any other joints. On examination of the right ankle joint, there was an ovoid swelling of 4x3cms located just below the lateral malleolus which was mobile, firm in consistency andnon trans-illuminant, suggestive of a lipomatous tumor.
Ankle range of motion was normal and no loose bodies or crepitus were palpable in the joint. Plain x-rays [antero-posterior and lateral views] of ankle joint showed a single calcified body measuring 1x1cms lying just underneath the tip of lateral malleolus (FIGURE-I). Rest of the ankle joint appeared normal except for the presence of ostrigonum but without evidence of degenerative changes. With this atypical radiological picture, we advised the patient for an MRI of the ankle, but due to financial constraints patient refused it. Regardless, we decided to proceed with excisional biopsy. A lipomatous swelling [6x5cms] was excised and beneath it was a solitary calcified body measuring 1x1cm lying outside the joint capsule (FIGURE-II). The wound was closed and the tissues were sent for Histo-Pathological Examination [HPE]. Post-operative days were uneventful. HPE reported as extra-synovial extra-articular synovial chondromatosis of the ankle (FIGURE-III). Before discharge, patient gave her consent for documenting this case for academic purpose. Unfortunately, patient never returned for follow-up after 3 months and the status of recurrence/ malignant transformations cannot be commented.

DISCUSSION

Primary synovial chondromatosis is a benign self-limiting lesion of unknown etiology that may recur locally. Even though, previously it was thought to be a metaplastic process of the synovial tissue, on the basis of currently known molecular abnormalities, it is now considered as a benign neoplastic disease (1). The extra-articular subtype typically arises from the synovium of the tendon sheath or bursa. The role of trauma in the development of this condition is uncertain, but history of trauma can mislead to the diagnosis of a trivial sprain. The condition is frequently seen in 3<sup>rd</sup> to 5<sup>th</sup> decade with a male preponderance, however when extraarticular disease is diagnosed
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in older patients [i.e., after the fifth decade], there is a female predominance [2:1 ratio] (2). Clinical manifestations in extra articular subtype can be subtle in the form of a painless mass or mild tenderness upon palpation with seldom limitation in range of motion. The classical radiological appearance of synovial chondromatosis is multiple, calcified bodies in juxta articular location. These calcifications frequently show a pathognomonic appearance of ring and arc pattern of mineralization. Sometimes, these individual loose bodies coalesce to form a giant single chondroma and become symptomatic. Neither of these two characteristic pictures were seen in the index case. Three differential diagnosis that could be made from the current clinico-radiological picture were: a calcified ganglion, fibrinous rice bodies [Tubercular arthropathy], and symptomatic os subfibulare. Ganglion and synovial cysts are the most common soft tissue lesions in the ankle and foot region and are usually associated with diseases like post-traumatic, inflammatory or degenerative joint diseases (3); moreover calcification in these cysts is not uncommon. Os subfibulare are separated ossicles at the tip of the lateral malleolus which can be another source of chronic ankle pain (4). Additional investigations like ultrasound, CT, and MRI further aid in diagnosing and choosing surgical approach. However, the final word on diagnosis could be made only by HPE. Microscopically, synovial osteochondromatosis is characterized by lobules of hyaline cartilage with variable degree of synovial proliferation or hyperplasia. The hyaline cartilage in primary synovial chondromatosis is often hypercellular with atypical histologic features, including multinucleation, nuclear crowding, nuclear enlargement and hyperchromasia, and mild myxoid changes, which would otherwise suggest a malignant cartilage neoplasm [grade 1 to grade 2 chondrosarcoma] (1). Associated foci of ossification is also noted in the cartilage; thereby the term ‘osteo’-chondromatosis. Secondary synovial chondromatosis can be distinguished from primary disease both radiologically [underlying articular disease and
fewer chondral bodies of variable size and shape] and pathologically [concentric rings of growth] (1).

Open surgical excision in the preferred treatment for this condition with prompt relief of the symptoms. Recurrence rate reported for extra articular subtype is highly varied, but most recurrences occur within 5 years of initial resection. The potential of malignant transformation into chondrosarcoma is seen in up to 5% of cases which further warrants a close post-operative follow-up (5).

CONCLUSION

Synovial osteochondromatosis is a rare cause for chronic ankle pain. Thorough clinical evaluation supported by radiological investigations aid in clinching the diagnosis. Surgical excision is the preferred modality of treatment. Long term follow-up is a must to rule out rare possibilities of recurrence or malignant transformation.

ACKNOWLEDGEMENT

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REFERENCES


### Table: Case reports on extra-articular synovial chondromatosis of the ankle joint

<table>
<thead>
<tr>
<th>Sl. no</th>
<th>Author/year</th>
<th>Age/sex</th>
<th>Presentation</th>
<th>Radiology</th>
<th>Treatment</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>S. B. Tibrewal et al <a href="6">1995</a></td>
<td>44/F</td>
<td>Painful mass, But pain-free movements</td>
<td>Bony erosion of tibio-fibular syndesmosis. CT revealed calcification in mass</td>
<td>Open Excision</td>
<td>2 years. No recurrence and asymptomatic</td>
</tr>
<tr>
<td>3.</td>
<td>B. Carpenter et al [2010] (8)</td>
<td>56/F</td>
<td>Painful dorsiflexion &amp; antero-medial mass</td>
<td>Single radio-opaque nodule. MRI done.</td>
<td>Open excision</td>
<td>9 months. Patient is asymptomatic</td>
</tr>
<tr>
<td>4.</td>
<td>S. Bahari et al [2012] (9)</td>
<td>53/F</td>
<td>Ankle sprain history, Pain &amp; swelling</td>
<td>Erosive lesion in ankle. MRI was required.</td>
<td>Open excision</td>
<td>1 year. Returned to normal activity</td>
</tr>
</tbody>
</table>
Fig. 1: Plain x-ray antero-posterior and lateral view of ankle joint
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Fig.2: Lipomatous swelling and solitary osteochondromatosis lying outside the joint.

Fig.3: Histopathology: 5X cartilaginous cap overlying trabeculae of bone.