CASE REPORT

Extra-articular Extra-synovial Solitary Osteochondromatosis of the Ankle

S Madi, S Vijayan, MA Naik, SK Rao

ABSTRACT

Synovial chondromatosis or osteochondromatosis is a benign neoplastic condition arising from synovial tissue of joints, tendon sheath and bursa. The commonly involved joints are the knee, hip, shoulder, elbow and ankle. According to the author's 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, osteochondromatosis, solitary, synovium.

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. The radiological picture is not only characteristic but almost diagnostic. There have been only four case reports to 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 to give 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 the subtle presentation.

CASE REPORT

A 49-year-old female without pre-existing co-morbidities presented to our foot and ankle clinic with complaints of dull aching pain of more than 3 years' duration in her right ankle. For the past 1 year, she also noticed a soft swelling over the lateral aspect of the ankle. She consulted a general practitioner who treated her with painkillers and herbal medicines, thereby diagnosing it as degenerative osteoarthritis of the ankle joint. The pain subsided with medications and rest but was aggravated by prolonged standing and walking on uneven ground. There was no history of trauma, constitutional symptoms

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

Sr. no	Author/year	Age/sex	Presentation	Radiology	Treatment	Follow-up
1.	Tibrewal and Iossifidis 1995 (6)	44/F	Painful mass, but pain-free movements	Bony erosion of tibiofibular syndesmosis. CT revealed calcification in mass	Open excision	2 years. No recurrence and asymptomatic
2.	Pathak *et al* 2006 (7)	32/M	Restricted dorsiflexion and locking episodes	Multiple loose bodies (intra-articular and extra-articular)	Arthroscopic synovectomy + loose body removal + open excision	1 year. Pain-free full range of ankle movement
3.	Carpenter and Clyde 2010 (8)	56/F	Painful dorsiflexion and antero-medial mass	Single radio-opaque nodule MRI done	Open excision	9 months. Patient is asymptomatic
4.	Bahari and McKenna 2012 (9)	53/F	Ankle sprain history, Pain and swelling	Erosive lesion in ankle. MRI was required	Open excision	1 year. Returned to normal activity

From: Department of Orthopaedics, Kasturba Medical College, Manipal, Manipal Academy of Higher Education, Karnataka 576104, India.

Correspondence: Dr S Vijayan, Department of Orthopaedics, Kasturba Medical College, Manipal, Manipal Academy of Higher Education, Karnataka 576104, India. Email: sandeep.vijayan18@gmail.com

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or involvement of any other joints. On examination of the right-ankle joint, there was an ovoid swelling of 4 × 3 cm located just below the lateral malleolus which was mobile, firm in consistency and non-transilluminant, which was suggestive of a lipomatous tumour. 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 the ankle joint showed a single calcified body measuring 1 × 1 cm lying just underneath the tip of the lateral malleolus (Fig. 1). The rest of the ankle joint appeared normal except for the presence of os trigonum but without evidence of degenerative changes. With this atypical radiological picture, we advised the patient to have a magnetic resonance imaging (MRI) of the ankle, but due to financial constraints the patient refused. Regardless, we decided to proceed with an excisional biopsy. A lipomatous swelling (6 × 5 cm) was excised and beneath it was a solitary calcified body measuring 1 × 1 cm lying outside the joint capsule (Fig. 2). The wound was closed and the excised tissues were sent for histopathological examination (HPE). Postoperative days were uneventful. Histopathological examination was reported as extra-synovial extra-articular synovial chondromatosis of the ankle (Fig. 3). Before discharge, the patient gave her consent for documenting this case for academic purpose. Unfortunately, the patient never returned for follow-up after 3 months and the status of recurrence/malignant transformations cannot be commented on.

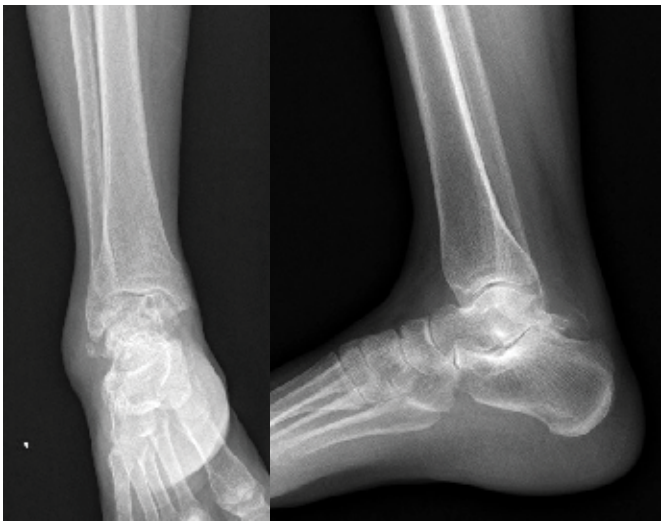

Fig. 1: Plain X-ray antero-posterior and lateral view of ankle joint.

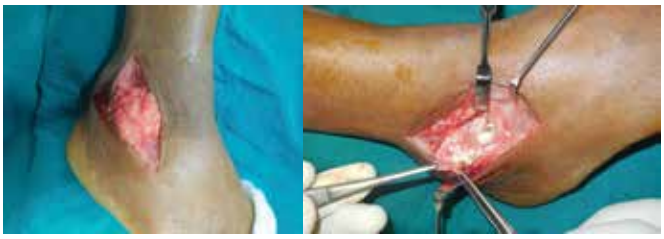

Fig. 2: Lipomatous swelling and solitary osteochondromatosis lying outside the joint.

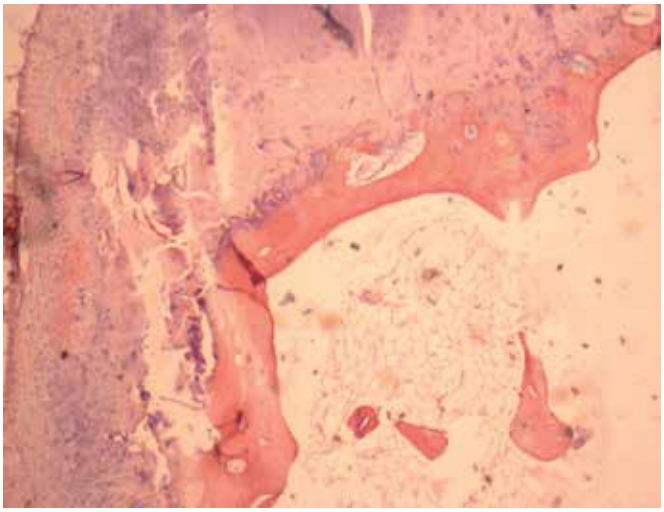

Fig. 3: Histopathology: 5× cartilaginous cap overlying trabeculae of bone.

DISCUSSION

Primary synovial chondromatosis is a benign self-limiting lesion of unknown aetiology 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 3rd to 5th decade with a male preponderance; however, when extra-articular disease is diagnosed in older patients (*ie*, after the 5th decade), there is a female predominance (2:1 ratio) (2). Clinical manifestations in the 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 the 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 diagnoses that could be made from the current clinicoradiological 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, computed tomography and MRI further aid in diagnosing and choosing the 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, and hence 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 is the preferred treatment for this condition with prompt relief of the symptoms. Recurrence rate reported for the 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 reinforces a close postoperative 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 necessary to rule out rare possibilities of recurrence or malignant transformation.

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