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Giant Extra-Articular Synovial Chondromatosis of the Ankle Joint – A Rare Case Report

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Learning Point of the Article:

Extraarticular swellings around the ankle joint may have synovial origin, though they are very rare.

Abstract

Introduction: Synovial chondromatosis (SC) commonly involves large joints such as the knee and hip with smaller joints being less frequently involved. Extra-articular involvement is very rare, and here, we are presenting the first case of giant extra-articular SC originating from the ankle joint.

Case Report: A 42-year-old male presented to the outpatient department with painless swelling over the lateral malleolus for 2 years. Diagnostic imaging suggested the involvement of the synovial lining with the swelling. The mass was excised and histopathology proved the diagnosis of SC.

Conclusion: Extra-articular involvement in SC has been mainly reported in the synovial sheath or bursae of the hand and foot, but they can involve ankle joint also. In recent times, there have been concerns about potential malignant transformation of these lesion to chondrosarcoma, diagnosing these lesions have become important.

Keywords: Ankle joint, Synovial chondromatosis, Extra-articular.

Introduction

Synovial chondromatosis (SC) is a disease of unknown etiology with a benign clinical course, characterized by the presence of metaplastic cartilaginous nodules (chondroma) within the synovial membrane [1]. These nodules can ossify, leading to extrusion from the synovium, leaving osteochondral loose bodies within the joint, or extra-articular soft tissues [2]. SC most commonly presents in the third to fourth decade of life with a male predilection and often involves large joints such as knee and hip with smaller joints being less frequently involved [3]. The number of chondroma varies anywhere from two or three to several dozen, but sometimes the chondromas coalesce or a single chondroma enlarges [4].

Edeiken et al., in 1994, first used the term giant SC for the synovial chondromas of more than 1 cm and occasionally reaching up to 20 cm of diameter. Extra-articular involvement is very rare and has been mainly reported in the synovial sheath or bursae of the hand and foot [3]. To the best of our knowledge, we are presenting the first case of giant extra-articular SC originating from the ankle joint.

Case Report

A 42-year-old Indian male presented to the outpatient department with chief complaints of nodular mass over lateral malleolus of the right ankle for 2 years (Fig. 1). The swelling was initially painless but later became painful while sleeping on the affected side of the ankle for the last 6 months. The patient had no history of trauma, past medical illness, or family history of similar illness. There were no vascular or neurologic abnormalities distal to the swelling. Local examination revealed a painless hard swelling and non-tender with irregular margins and was not fixed to the overlying skin. There was no restriction of the ankle movements and was painless in all ranges of motion.

A plain radiograph revealed a large spiculated calcified mass

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Author's Photo Gallery


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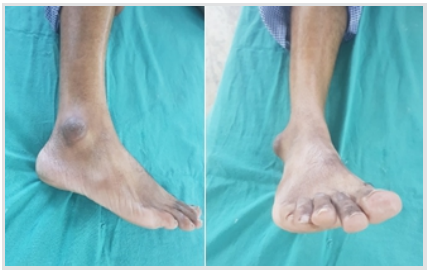

Figure 1: Pre-operative photographs showing the swelling over the lateral side of the ankle.

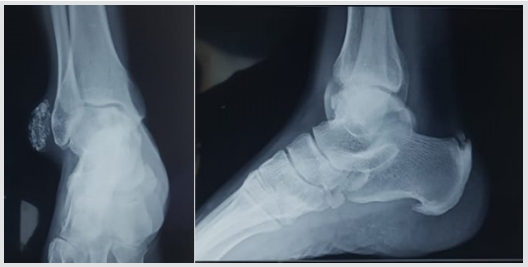

Figure 2: Pre-operative anteroposterior and lateral X-ray showing calcified mass over the lateral malleolus.

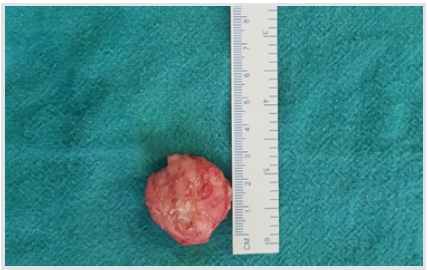

Figure 3: The macroscopical aspect of the excised mass over the lateral malleolus.

over the lateral malleolus of the right ankle (Fig. 2). There was no visible osseous destruction or scalloping. Magnetic resonance imaging (MRI) showed isointensity on T1-weighted image and T2-weighted image showed high signal intensity consistent with calcification.

Based on the clinicoradiological findings, surgical excision of the mass was planned under regional anesthesia. The lesion was completely excised along with its pseudo capsule, as there was no adherence to any of the surrounding bony or soft-tissue structures. There was no synovial thickening or proliferation; therefore, synovectomy was not performed. Since there was no articular involvement, weight-bearing was allowed as soon as the pain subsided.

The gross pathological examination revealed a single giant chondroma with an irregular margin. The irregular tissue was 3×2×1 cm in size (Fig. 3). Histopathological examination confirmed the diagnosis of SC (Fig. 4).

Discussion

SC is a condition with a benign course that usually occurs among individuals over 40 years of age, with a male predilection [5]. SC is characterized by metaplastic changes in the synovial membrane of the joints, tendons, and bursae, leading to the formation of multiple cartilaginous nodules. Involvement is typically monoarticular, with large joints being affected most frequently. The knee joint is most commonly affected and accounts for 60–70% of cases, and the shoulder, elbow, and hip are the next frequent sites [6]. Extra-articular involvement of the ankle joint by Giant SC is very rare. A review of English literature has not shown any article with a similar presentation.

The etiology of SC is not known, but certain data have suggested a neoplastic origin with an abnormality of chromosome 6 [2]. SC can be classified into primary and secondary forms. The primary type occurs in an otherwise normal joint and is associated with benign reactive metaplasia of the synovial membrane. This form is usually progressive, more likely to recur, and its long-term presence is associated with severe degenerative joint disease [7]. Milgram et al. classified the primary SC into three phases. They described Phase I as active synovial diseases with no loose bodies. Phase II is a transitional phase with active intrasynovial pathologic tissue with loose bodies. Phase III encompasses several free osteochondral bodies with intrasynovial disease. Secondary SC is associated with joint abnormalities, such as degenerative arthritis, trauma, neuropathic arthropathy, inflammatory arthropathy, tuberculosis, and avascular necrosis [8,9].

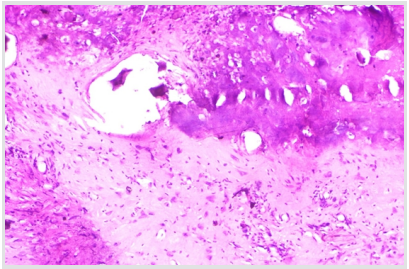

Figure 4: The microscopical aspect of the swelling (hematoxylin and eosin image, ×ss400) showing atypical chondrocytes with the presence of ossification.

Clinically, these patients present with pain, swelling, stiffness, and/or growing palpable mass on affected joints, and diagnosis of extra-articular SC may be difficult or delayed until surgical treatment, as symptoms are vague and non-specific. In recent times, there have been concerns about potential malignant transformation of these lesion to chondrosarcoma, diagnosing these lesions have become important. Monesteir et al. concluded that in the case of known primary SC, if there is the rapid deterioration of the clinical symptoms, transformation to synovial sarcoma should be suspected [7]. Bojanic et al. reported malignant transformation in 17–25% of all cases, and this transformation is closely connected to the recurrence rate. They also concluded that malignant transformation usually occurs many years after operative treatment, and therefore, long-term follow-up is necessary for these lesions [10].

Kim et al. reported a case of giant extra-articular SC originating from the sinus tarsi in a 32-year-old male. The swelling was painful and had affected the daily activities. Diagnostic imaging suggested the diagnosis of the giant extra-articular SC. The bony mass was excised, and the patient was doing all the activities of daily living without pain at 2 years follow-up [11].

Fornaciari et al. reported a case of intra-articular giant osteochondroma of the ankle joint in a 17-year-old male. The patient was managed by complete excision through a posteromedial arthrotomy of the ankle joint. After 2 years, the patient was asymptomatic and was doing a recreational activity on all types of surfaces [12].

Plain radiographs are usually the first-line investigation. Extra-

articular SC is difficult to diagnose in the early stages. The radiological appearance depends on the average time of diagnosis from the onset of symptoms and calcification of the cartilaginous nodule. If the nodule is adequately calcified, a plain radiograph may give a clue to the diagnosis. A computed tomography scan clearly depicts the calcified nodules and documents joint destruction and extrinsic joint erosion [7, 13]. MRI scans are usually needed in the early phase of SC, before the occurrence of calcification or ossification of nodules. MRI is the best modality to evaluate the extent of the lesion, extrinsic bone erosion, and marrow invasion [7].

The goal of treatment is to relieve pain, removal of the loose bodies, regain movement of the joint, and prevent the progression of osteoarthritis and chondral damage. Excision of the loose bodies and extensive synovectomy regardless of extra-articular and intra-articular chondromatosis constitutes the standard line of management of these lesions [12]. Surgical management is usually by open method or arthroscopy assisted. Post-surgery, the recurrence rate is 3–23% and is thought to be due to remnant active synovium after synovectomy or the presence of the stimulus causing metaplasia [7]. In our case, since the mass was extra-articular, the only excision of the mass was performed and arthrotomy was not done.

Following the surgical excision, the patient was symptom-free at the 3-months follow-up. The disease usually has a benign course, but the chances of malignant transformation also have to be kept in mind; therefore, long-term follow-up is necessary for these cases.

Conclusion

SC is usually confined within the joint and extra-articular involvement is very rare. Extra-articular involvement has been mainly reported in the synovial sheath or bursae of the hand and foot, but they can involve ankle joint also. In recent times, there have been concerns about potential malignant transformation of these lesion to chondrosarcoma, diagnosing these lesions have become important.

Clinical Message

Swellings over the joints can have a synovial origin. Therefore, histopathological examination is necessary in all cases to confirm the diagnosis. SC should be followed-up for a long duration due to the risk of malignant transformation.

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