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"Synovial chondromatosis involving small joints like ankle: A rare case report"

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Abstract:
Background*: Primary synovial chondromatosis* ***(****Reichel syndrome****),*** *is a benign mono-articular disorder of unknown origin characterized metaplasia and proliferation of synovial tissue resulting in multiple intra-articular cartilaginous loose bodies of relatively similar in size usually involving larger joints commonly found in men of middle age of third to fourth decade of life.*
Case characteristic: *A 40 year male presented with dull aching pain in the ankle joint left side since last one year which is progressively increasing in intensity along with swelling of the left ankle more towards the anterior aspect with restriction of movement. On examination, there was synovial hypotrophy and crepitation inside the joints with ROM 10 degree dorsiflexion with15 degree planter flexion .X-ray of the ankle showing multiple loose body mostly located the anterior part the ankle. MRI showing multiple intra-articular mass iso-intense to muscle in T1W and hyper-intense to muscle on T2WA containing multiple foci of low signal.*
Outcome: *The patient underwent open arthrotomy with debridement and removal of the loose body with synovectomy, followed for one and half years without any recurrence now walking comfortably with full range of motion and regained his previous daily activity level.*
Conclusion: *Primary synovial chondromatosis of the ankle which is an unusual rare presentation.In a patient with long standing history of pain swelling and crepitation even in a small joint clinical diagnosis of primery synovial chondromatosis should be considered and further more thorough radiological evaluation and histopathological evaluation will add to the diagnosis*

Keywords: *Synovial chondromatosis, Loose bodies, Ankle joint, Open arthrotomy.*

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I. Introduction

Primary synovial chondromatosis **(**Reichel syndrome)[1]**,** is a benign mono-articular disorder of unknown origin characterized metaplasia and proliferation of synovial tissue resulting in multiple intra-articular cartilaginous loose bodies of relatively similar in size usually involving larger joints of body most commonly involving knee joint , other joints being hip , shoulder[2,3,4] . Involvement of ankle is relatively rare[5,6]. These loose bodies under high pressure may coalesce with each other and become a large calcified body inside the joint. Commonly found in men of middle age of third to fifth decade of life, incidence being twice in males as compared to females. Abnormalities of chromosome 6 may be associated with this condition. Pain, swelling, restriction of movement are being the common presentations. We are presenting a rare case of synovial chondromatosis involving ankle joint treated by open arthrotomy and regaining back to his previous functional status.

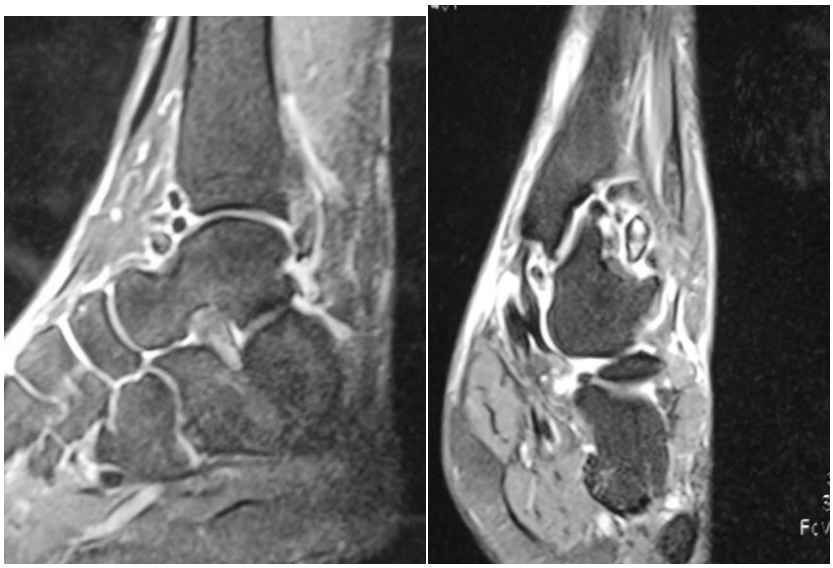
II. Case Report

A 40 year male presented with dull aching pain in the ankle joint left side since last one year which is progressively increasing in intensity along with swelling of the left ankle more towards the anterior aspect with restriction of movement along with crepitation. The pain was increasing on walking and subsiding on rest. There was no history of the trauma to the ankle, fever, previous swelling or any other joint involvement.

On physical examination, there is no local raise of temperature or tenderness. There was synovial hypotrophy and crepitation inside the joints with ROM 10 degree dorsi flexion and 15 degree planter flexion. There was no instability of ankle or any distal neurovascular deficit. On further evaluation, all the laboratory, hematological and biochemical parameters found to be within normal range. X-ray of the ankle showed multiple

calcified mass of loose bodies located mostly anterior aspect of the ankle. On further evaluation with MRI multiple intra-articular mass iso-intense to muscle in T1W and hyper-intense to muscle on T2WA containing multiple foci of low signal intensity with minimal collection seen in the tendon sheath suggestive of synovial chondromytosis 6. Other bones distal tibia and fibula including malleoli appeared normal. The patient had undergone open arthrotomy of the left ankle with anterior approach to the ankle joint. Intra-operatively, we found multiple intra articular loose bodies which were homogeneous in appearance with size of 2 to 8mm, smooth surface accounting more than 60 in number. Ankle was thoroughly irrigated with normal saline and partial synovectomy was done. Post operatively compression bandage was applied and allowed partial weight bearing for 2 weeks. Then full weight bearing was allowed afterward. Post operative radiograph was obtained showing no intra articular loose body. The specimen was sent for histo-pathological evaluation confirms the diagnosis of primary synovial chondomatosis. After one and half year of follow up, patient is walking comfortably with full range of motion. He has regained his previous daily activity level.

"Figure .1": Lateral radiograph of ankle showing multiple loose bodies, mostly in the anterior aspect of ankle joint



"Figure.2": Magnetic resonance imaging (Sagittal and coronal T2image) showing multiple foci of low signal intensity with minimal collection seen in the tendon sheath suggestive of synovial chondromytosis

"Figure.3": Intra operative picture showing multiple pearl like bodies

III. Discussion

Primary synovial chondromatosis is a benign mono-articular disorder of unknown origin characterized metaplasia and proliferation of synovial tissue resulting in multiple intra-articular cartilaginous loose bodies of relatively similar in size usually involving larger joints of body; most commonly involving knee joint (60 - 70%)[8] . Other joints being hip, shoulder and elbow. Involvement of ankle and foot are rare. Only few cases of synovial chondromatosis with involvement of foot and ankle have been reported[9]. The exact etiopathogenesis of synovial chondromatosis is still unknown. Milgram, et.al. classified the disease process into three distinct stages[10, 11]. In first stage, synovial lining undergoes cartilaginous metaplasia. In the second stage, nodules begin to detach from the synovium and appear as intra articular lose bodies and patient becomes symptomatic in this stage. In the third stage, multiple loose bodies can be observed within the joint cavities with no visible intra synovial bodies indicating that activity in the synovium has settled. The intra articular loose bodied have the tendency to unite among themselves and calcify. Clinically patients with synovial chondromatosis usually present with pain swelling, stiffness of the joint and/or a mass inside the joints. Most of them have long standing clinical history before an accurate diagnosis is made. Malignant degeneration in few cases of primary chondromatosis has been reported[12]. In 70-90% of cases with clinical features along with radiological evaluation points towards diagnosis. Multiple intra articular calcified bodies with smooth, round and of variable sizes within the joint capsule are the findings with diagnostic significance. Although increase in the soft tissue density around the affected joints can be noticed in x-ray, MRI can clearly demonstrate and differentiate calcification and ossification.

Early detection of the disease, confirmation of the diagnosis and commencement of the treatment should be the goal through which we can regain the range of movement with symptomatic improvement of the patient and can prevent further development of secondary osteoarthritis by removal of the loose bodies and thorough debridement of the joint. Treatment of choice is either open or arthroscopic debridement with or without synovectomy. Patients presenting earlier in the disease process stage 1 and stage 2 , with active synovitis and synovial thickening , synovectomy is indicate in order to prevent recurrence of the disease . But in later stages as synovitis subsides, in those cases synovectomy is not indicated. In 3 to 23% of cases recurrence may occur and is thought to be after synovectomy with active synovium remaining or presence of stimulus which cause the metaplasia[13]. In our case, the patient underwent open arthrotomy with debridement and removal of the loose body with synovectomy. Patient was followed for one and half years without any recurrence. Although some studies emphasise on the importance of arthroscopic debridement of the joint with removal of loose bodies as better as compared to open arthrotomy but this may not be applicable to smaller joints like ankle[7]. Considering ankle being a smaller joint in comparison to knee and shoulder, open arthrotomy is the technique of choice for thorough debridement and complete removal of loose bodies along with synovectomy as it is a better, easier and less time consuming with good exposure.

IV. Conclusion

Primary synovial chondromatosis of the ankle is an unusual rare presentation. In a patient with long standing history of pain swelling and crepitation even in a small joint clinical diagnosis of primary synovial chondromatosis should be considered and further more thorough radiological evaluation and histopathological evaluation will add to the diagnosis. Goal of the treatment being through debridement, complete removal of the loose bodies with or without synovectomy either through open arthrotomy or arthroscopically.

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