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Case-based Review

Retrospective analysis of the clinical and radiological profile of few cases of synovial osteochondromatosis with a literature review

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Abstract

Synovial osteochondromatosis (SOC) is a rare benign neoplasm. It is caused by the metaplasia of the synovium into the bone. The knee, hip, elbow, shoulder, and ankle are the common sites. Our objective was to retrospectively assess the clinical and radiological features of a few cases of SOC resembling some rheumatological conditions and describe it along with a literature review. A retrospective analysis of the clinical and radiological features of a few cases of SOC mimicking some rheumatological conditions was performed. There were 4 cases: 3 adult men and a young boy. Three cases presented in the elbow and 1 case in the hip. There was a case mimicking osteoarthritis of the elbow in a young individual, another case presented like myositis ossificans of the elbow, and another case was similar to neuropathic arthritis of the elbow. The case in the hip presented as painful limping in the child. SOC can sometimes mimic various conditions affecting the synovial joints. A careful evaluation of the clinical and radiological features can be helpful in the correct diagnosis.

Keywords: Synovial osteochondromatosis, myositis ossificans, neuropathic joint, painful limping child

Introduction

Synovial osteochondromatosis (SOC) is a rare entity. It is caused by metaplasia of the synovia into the bony tissue. It is common in men in the fourth or fifth decade of life. Any synovial joint can be affected. It can be seen in the tenosynovium. The knee is the most common site. It usually affects a single joint. The most common presentations are pain and limitation of movements. The treatment includes removal of loose bodies with synovectomy. However, complete removal is impossible in most cases; hence, recurrence is common (1).

In this retrospective study, we discuss the clinical and radiological features of 4 cases of SOC mimicking other rheumatological conditions and how to differentiate it from other conditions with a literature review.

Case Presentation

A 41-year-old man, a manual laborer, presented with pain and swelling of the right elbow for approximately 1 year. The pain was gradual in onset and progression. After about 6 months, he noticed swelling of the elbow with limitation of movements. At the time of presentation, there was a boggy swelling of the right elbow with tenderness. There was a fixed flexion deformity of 30° with further flexion up to 80°. Hemogram was normal. No elevation of erythrocyte sedimentation rate and C-reactive protein levels was observed. The radiograph showed widening of the soft tissue shadow with multiple loose bodies. There was narrowing of the joint space with subchondral sclerosis and osteophyte formation (Figure 1). Magnetic resonance imaging (MRI) showed multiple rounded and oval loose bodies in the distal part of the humerus, intra-articular with effusion. There were features of early degenerative changes (Figures 2 and 3). His diagnosis was SOC of the right elbow. He was treated by synovectomy and loose body removal. Postoperatively, he got symptomatic relief. He got more than 100° of flexion after 3 years (Figures 4 and 5).

The second case was of a 35-year-old man who presented with pain and swelling of the left elbow for approximately 2 years. His symptoms started after minor trauma. He was more concerned about his progressive loss of range of movements. On examination, he had a stiff elbow with swelling. Radiographs showed

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a long calcific intra-articular mass bridging the coronoid and anterior humerus with loose bodies posteriorly in the joint. There were no degenerative changes (Figure 6). Synovectomy and loose body removal did not improve his range of movements.

The third case was of a 52-year-old man with almost similar presentation. However, there were multiple loose bodies and synovial thickening. Radiographs also showed degenerative changes (Figure 7). Neuropathic elbow was the differential diagnosis. His blood glucose level was normal, and there was no clinical evidence of motor or sensory involvement. MRI of the cervical spine was normal.

The fourth case was of a 15-year-old boy who presented with a gradual onset of pain of the right hip for 1 year. Thereafter, he developed limping. There was no history of trauma or constitutional symptoms. He had an antalgic gait with tenderness over the right hip. The movements in all directions were restricted. Maximum restriction was in the internal and external rotations. All routine blood investigations were normal. The X-ray image showed capsular distension with multiple loose bodies in the right hip. There was no loss of joint space or degeneration (Figure 8). A contrast-enhanced MRI scan showed a thickened capsule with effusion and mild erosions in the acetabulum with multiple loose bodies in the joint (Figures 9 and 10). It was a case of SOC of the hip. He was treated by synovectomy and loose body removal through an anterior approach (Figures 11 and 12). He got relieved of the pain, and there was improvement in the movement. We could not remove all the loose bodies (Figure 13).

Main Points

- Synovial osteochondromatosis (SOC) is a rare benign neoplasm affecting the synovial joint. Pain and limitation of movements are the common clinical features with intra-articular loose bodies on a radiogram.
- SOC can rarely present early-onset osteoarthritis, myositis ossificans, neuropathic joint, or painful limping.
- SOC can be included in the differential diagnosis of osteoarthritis of synovial joints in young individuals. Ankylosis of the elbow with bony bridges can be due to SOC. Degenerative changes with multiple loose bodies in adults can be due to SOC. Atypical presentation of SOC can mimic other conditions in a child with a painful limp.

Informed consent was obtained from the patients.

Discussion

In this series, we described 4 cases of SOC. The first case was of a young man who presented with features of arthritis in the elbow. There was a painful restriction of the elbow movements with flexion deformity. Tuberculosis, post-traumatic arthritis, hemophilic arthritis, and ochronosis are the common causes of painful chronic arthritis in young men. Primary osteoarthritis of the elbow joint is rare. There was no history of trauma, constitutional symptoms, or pigmentation of the pinna or nose. Radiographic findings of osteoarthritis of the elbow show narrowing of ulnotrochlear joint, osteophytes in the olecranon and coronoid, and loose bodies. In tuberculosis, there will be osteoporosis and global narrowing of the joint space, and periarticular erosions will be present. Radiogram in hemophilic arthropathy shows enlargement of the epiphysis and osteoporosis. Absence of osteoporosis helps us to rule out

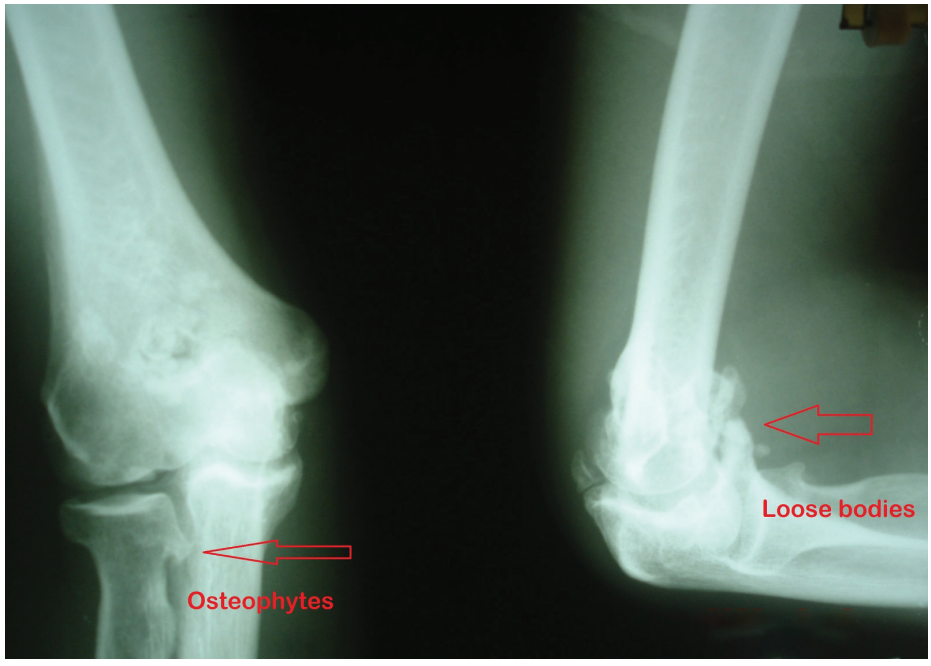

Figure 1. X-ray image of the right elbow. Anteroposterior and a lateral view showing multiple loose bodies, narrowing of the joint space, subchondral sclerosis, and osteophyte formation.

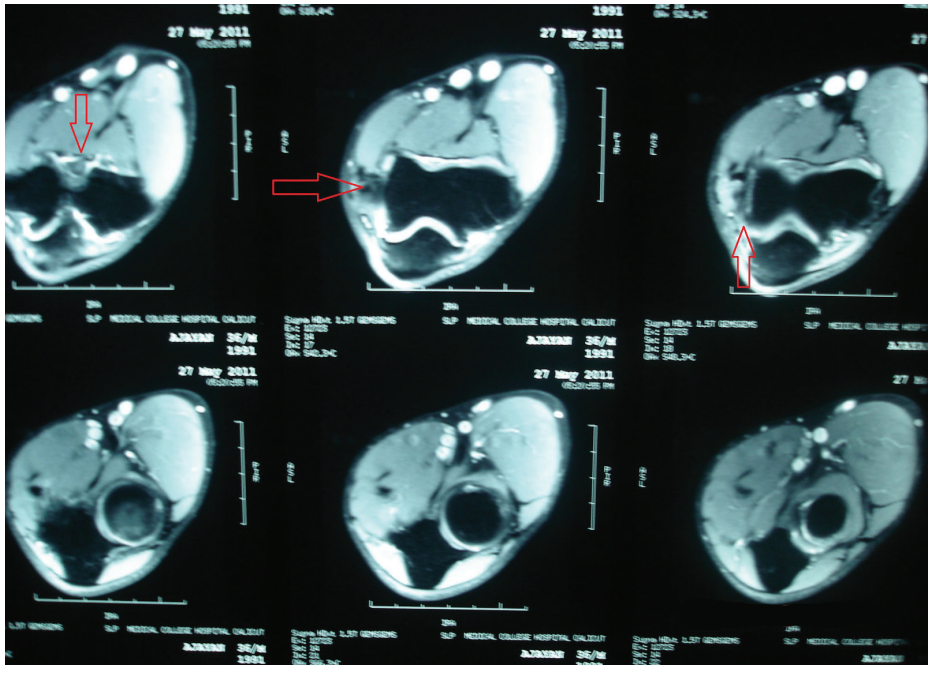

Figure 2. Magnetic resonance imaging of the right elbow coronal section. T1-weighted images showing multiple hypointense loose bodies.

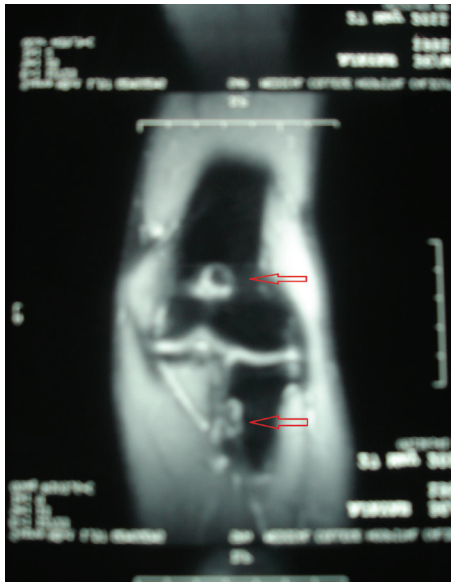

Figure 3. Magnetic resonance imaging of the right elbow sagittal section. T2-weighted images showing multiple hyperintense loose bodies.

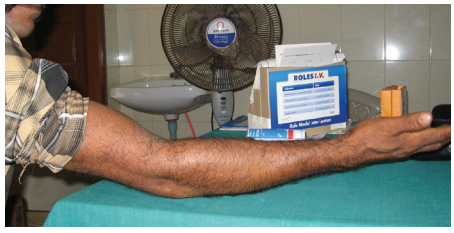

Figure 4. Postoperative function. The extension is about 20° short of normal.

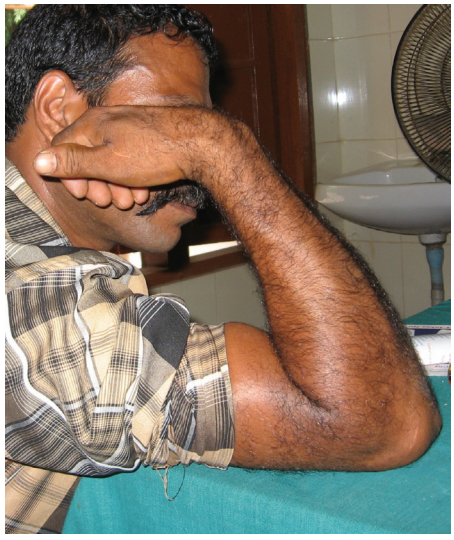

Figure 5. Postoperative function. Postoperative flexion of the right elbow is about 110°.

tuberculosis and hemophilic arthritis. There was no history of trauma and no evidence of malunion in the elbow. Thus, we came to the diagnosis of SOC in this case. Therefore, SOC can be considered as a differential diagnosis of osteoarthritis in young patients (2-4).

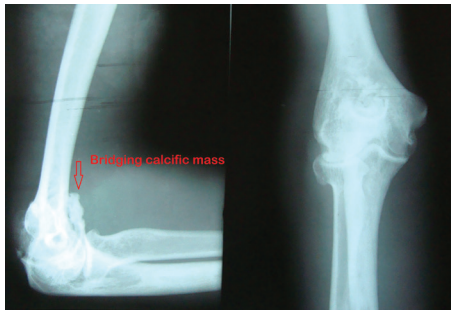

Figure 6. X-ray image of the left elbow. Anteroposterior and a lateral view showing a calcific mass bridging the coronoid process of the ulna to the anterior cortex of humerus. The absence of trabeculae and density of the lesion lower to cortical density helps to differentiate it from an ossific lesion.

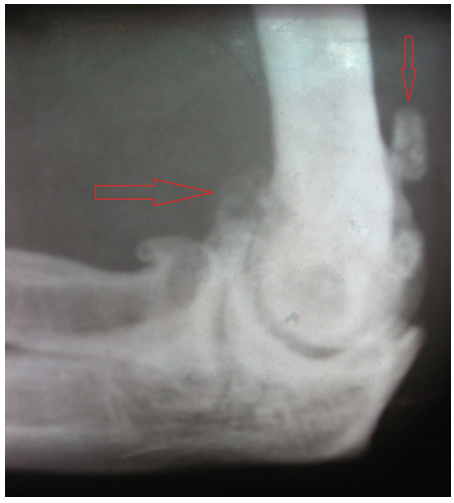

Figure 7. X-ray of the elbow joint. Lateral view showing multiple loose bodies and degenerative changes, mimicking the diagnosis of the neuropathic elbow joint.

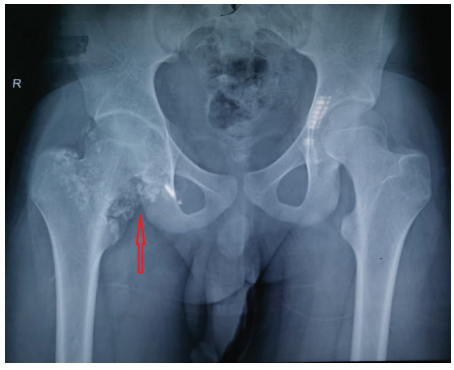

Figure 8. X-ray of the pelvis with both hips. There is distension of the joint with multiple loose bodies of varying sizes in the right hip joint. There are no features of degeneration.

Myositis ossificans is a benign condition. It is caused by metaplasia of the skeletal muscle into chondrocytes and osteocytes. There are 3 stages in the formation of myositis: early, intermediate, and late stages. Pain is very evident in the early stages, which subsides, and stiffness is the major problem later. The maturation of myositis occurs by 9 months when its borders become smooth and radiologically indistinguishable from the mature bone. It can be difficult to differentiate it from osteosarcoma in the early stages. Typically, myositis shows maturation in the periphery of the lesion with central immature bone in histology (Ackerman zones). A radiogram or computed tomography (CT) scan can demonstrate this peripheral maturation (5). In our case, there was stiffness of the elbow with deformity. The X-ray showed an anterior calcific lesion bridging the anterior humerus and coronoid. Despite the 2-year history, the borders of the lesions were irregular and immature. The absence of trabeculae in the mass helped us to differentiate it from an ossified lesion. We think soft tissue contracture due to deformity may be the reason for the lack of improvement in the range of movements after treatment. Therefore, radiological evaluation is important for differentiating SOC from myositis ossificans when presenting as a stiff elbow with calcified mass.

Neuropathic arthritis of the nonweight-bearing joints of the upper limb is rare. The most common causes are syringomyelia and leprosy. Osseous fragmentation and debris are very common and may confuse with the tumor matrix (6). Neuropathic arthritis of the elbow is very rare. One-third of patients present with pain. Although hypertrophic and atrophic changes can occur in the neuropathic joint, hypertrophic changes predominate in the elbow. Usually, the normal elbow architecture is lost, with osteophyte formation and heterotropic bone formation, and loose bodies are seen. Our patient presented with synovial hypertrophy, pain with degenerative changes, and loose bodies in the elbow radiogram. The neuropathic joint was a possibility we considered in this case. However, the absence of a predisposing factor, lack of joint disorganization, and instability were the points against it being a neuropathic joint (7).

There are many causes of a painful limping in a child. Legg-Calve-Perthes disease, slipped capital femoral epiphysis, infective arthritis, and juvenile chronic arthritis are common in adolescents. All of them present with pain and limitation of movements. Abduction and internal rotation are the common restricted movements. Osteochondritis dissecans and osteochondral fractures are the common causes of osteocartilaginous loose bodies in the hip in children. SOC can produce multiple loose bodies in the hip. Compared with other sites, SOC of the hip joint affects young individuals (8, 9). In our patients, all movements were restricted,

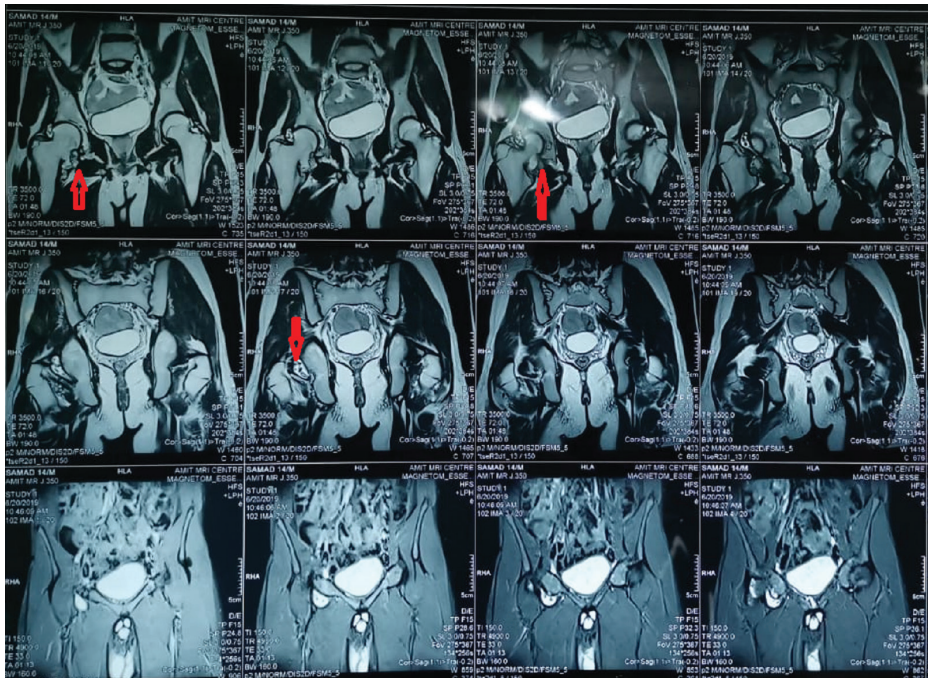

Figure 9. Magnetic resonance imaging showing coronal section of the pelvis. Loose bodies that are hypointense in T1-weighted images and hyperintense in T2-weighted images in the right hip.

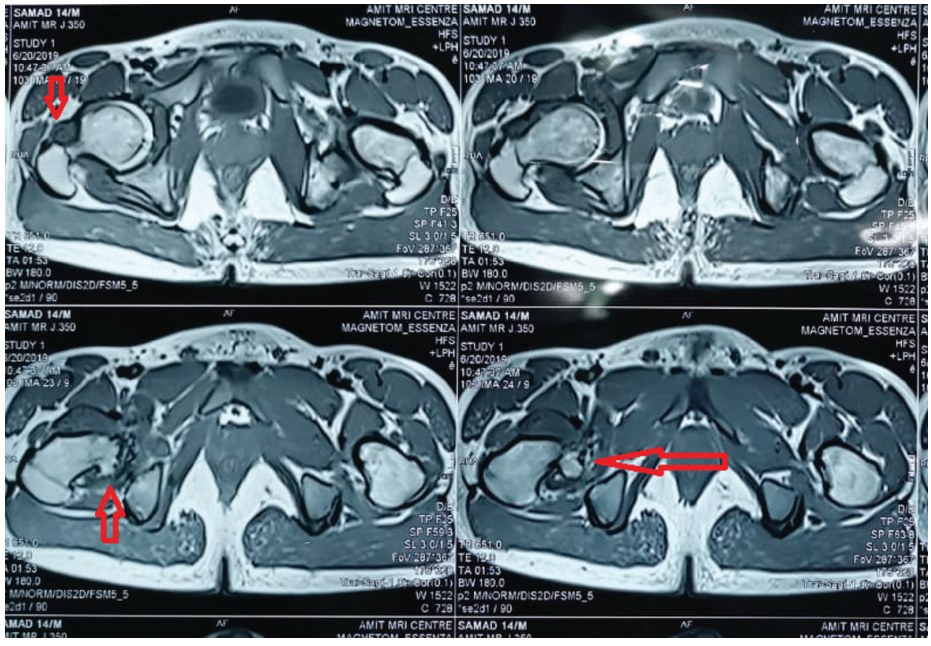

Figure 10. Transverse sections of magnetic resonance imaging showing multiple loose bodies in the right hip.

especially rotations. Clinically, owing to severe restriction of movements, we considered the possibility of idiopathic chondrolysis of the hip joint. However, typical deformity of flexion, abduction, and external rotation were not present. The X-ray shows osteoporosis and global narrowing of the joint space. In our case, there was no osteoporosis and narrowing of the joint space; instead, there were multiple loose bodies in the joint (10). There can be a joint space widening of the hip in SOC. There are studies regarding subluxation of the hip due to SOC. The extra-articular spread can occur in the hip. It can be treated by either open or arthroscopic mode (11-13). Therefore, SOC can rarely present in a child with a painful limp.

Primary SOC is also known as Reichel-Jones-Henderson syndrome. In 1558, Ambrose Paré first reported this condition. Thereafter, Laennec reported that loose bodies arise from subsynovial tissues (in 1813). In

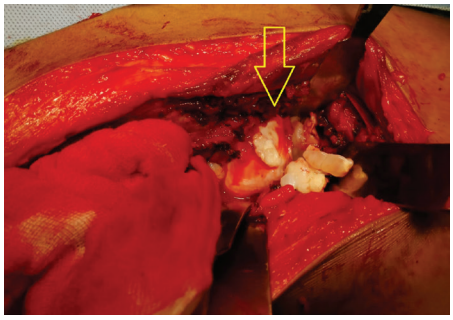

Figure 11. Intraoperative photograph. Right hip is opened through an ilioinguinal approach showing multiple cartilaginous loose bodies inside the joint.

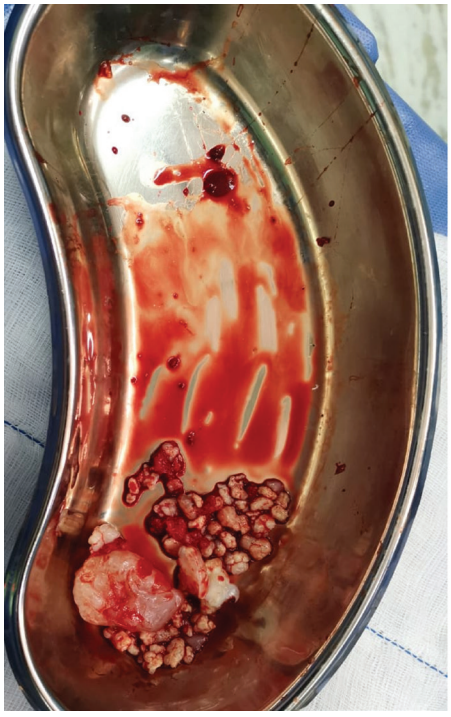

Figure 12. Loose bodies. Photograph showing the removed loose bodies from the right hip.

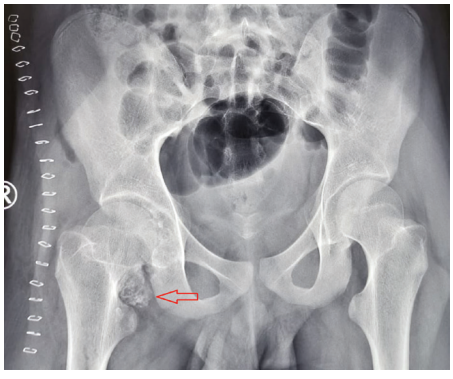

Figure 13. Postoperative X-ray image. Immediate postoperative radiograph showing incomplete removal of the loose bodies from the right hip.

1918, Henderson first reported SOC in the elbow joint. Pain and loss of extension are the most common symptoms. Loss of extension

may be due to pain, effusion, ulnar nerve impingement, or degenerative changes. In some cases, both flexion and extension will be lost. The exact mechanism of degenerative arthritis in SOC is unknown (14). In some cases, SOC is self-limiting and undergoes spontaneous resolution. Symptoms may be present for a long time before incapacitating the joint functions. Loose bodies formed by metaplasia of the synovium may either calcify or ossify; therefore, osteochondromatosis is a misnomer. The cause of metaplasia is unknown, but trauma is thought to be one (15). There are studies about SOC of the elbow joint causing ulnar nerve and median nerve palsy (16).

Primary SOC is now considered a benign neoplastic condition. It is characterized by the proliferation of the chondroid nodules in the synovium. There are 3 stages in the pathogenesis. In the initial stage, there is synovial hypertrophy and metaplasia into the cartilage. In the next stage, loose bodies get separated from the synovium and lie in the joint. In the final stage, the synovial proliferation stops, but the loose bodies increase in size, taking nutrition from the synovial fluid (17). There is an extremely low chance of transformation into chondrosarcoma (18). SOC usually affects single large synovial joints such as knee, hip, elbow, shoulder, and ankle. Smaller joints such as metacarpophalangeal, metatarsophalangeal, tibiofibular, and inferior radioulnar joints, are also affected. Radiography is the most common mode of diagnosis. Atypical presentations such as isolated synovitis can occur rarely (19). SOC can present with symptoms and signs that can masquerade as arthritis (20). There is a study on intra-articular melorheostosis of the knee mimicking SOC (21).

The routine blood investigations and serum calcium, phosphorus, and alkaline phosphatase levels are normal. The X-ray images show abnormalities in 70% of cases. In the initial stages, capsular distension with juxta-articular osteopenia is seen. Thereafter, multiple loose bodies are distributed intra-articularly and in the bursa and tendon sheaths; periarticular erosions (apple core sign) and features of degeneration are seen. Most cases can be diagnosed with radiographs alone (22). The MRI scan shows effusion and intra-articular loose bodies, which are hypointense in the T1-weighted images and hyperintense in the T2-weighted images. Kramer et al. (22) described 3 types of loose bodies in MRI: type A (chondromal), unmineralized loose body, difficult to distinguish with synovium; type B (osteochondral), in the majority due to mineralization; and type C, ossified loose bodies. These types correspond to pathological stages of diseases (23). An ultrasound scan shows that avascular masses surrounded by fluid nodules are hyperechoic. Dynamic ultrasonogram is also helpful. A CT scan identifies calcific loose bodies. Arthrogram can be used rarely (24). Diagnosis is confirmed by microscopy. On hematoxylin and eosin staining, thick villous formation in the synovium with islands of clustered chondrocytes is seen (25).

Differential diagnosis of SOC includes tuberculous arthritis, osteoarthritis, pigmented villonodular synovitis, gout, neuropathic joint, synovial sarcoma, and myositis ossificans. The ossific loose bodies can be differentiated from the calcific ones by looking at their density in the X-ray image, which is equal to cortical bone density in the ossified loose bodies. The presence of trabeculae in the loose body is another feature of an ossified loose body (25).

Inflammation and pain can be reduced using nonsteroidal anti-inflammatory drugs or ultrasound or other heat modalities. Mechanical symptoms can be tackled only with surgery. An open or arthroscopic synovectomy with the removal of the loose body is performed. There is a high rate of recurrence after surgery (0%-22%). Incomplete removal of the synovium and loose bodies are the reasons for recurrence (11).

SOC can mimic various conditions affecting the synovial joints. A careful evaluation of the clinical and radiological features can be helpful in the correct diagnosis.

Informed Consent: Informed consent was obtained from the patients.

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