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Primary Synovial Chondromatosis of Elbow: A Bowl Full of Tumors

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Abstract

Synovial osteochondromatosis is a disease of the synovium characterized by multiple cartilaginous bodies formation. Being characteristically monarticular it commonly involves the knee joint. Despite frequent mention of elbow involvement in various texts and articles, we wish to review the clinical, radiologic, and histologic features by presenting a case in an elderly male. By correct preoperative diagnosis and complete excision, full range of motion can be achieved in an relatively unforgivable elbow joint even with a delayed presentation.

Keywords: Loose bodies, Elbow, Synovial

Introduction

Synovial chondromatosis is a rare benign condition involving the synovial lining of joints, synovial sheaths, and bursae around the joints. The synovium undergoes metaplasia and gets converted into the cartilage which gets detached to become a loose body. Knee, hip, shoulder, ankle, elbow, and wrist are the most common joints involved. Smaller joints have also been involved, including distal radioulnar, tibiofibular, metacarpophalangeal, and metatarsophalangeal joint [1]. Bursae located around the joints are also rarely involved. Individuals usually belong to Third to fifth decade of life. Pain, swelling, and restriction of joint movements are most common presentations. Management is mainly surgical, either open or arthroscopic. We present a case of primary synovial chondromatosis in an elderly individual with a delayed presentation.

Case Report

64-year-old male patient presented to us with complaints of pain and swelling over the left elbow for 2 years. On presentation his main concern was pain and a sense of locking of the elbow. Pain aggravated on movement and is relieved with rest. There was no history of fall. The patient had not consulted any doctor for these complaints for the last two years. Local examination showed 4×3 cm oval-shaped swelling present over the posteromedial aspect of the elbow. Bilateral medial condyle prominence noted. On palpation, 5×4 cm multiple oval-shaped firm swelling present over the posteromedial aspect, mobile with the irregular edge with minimal tenderness. Range of movements of left elbow was 20 degrees with extension block with painful elbow flexion beyond 70 degrees with soft end point associated with crepitus and no instability. The patient was advised to get a routine blood investigation and X-ray of the elbow. Blood values were within normal range, and X-ray showed multiple stippled calcification-based loose bodies over the left elbow (Figure 1 A & B).

Based on the clinico-radiological finding, open surgery was planned. Through the Medial approach to the elbow done (Hotchkiss) multiple loose bodies (Figure 2) of varying size with the most prominent one measure 2×1×1 cm retrieved from anterior, medial, and posterior aspect along with a section of thickened synovial tissue which was sent for biopsy. The range of elbow movement was assessed intraoperatively, and elbow 0-120 degrees of range of motion was achieved. Under fluoroscopy, complete removal of loose bodies was confirmed.

Histopathology showed synovial tissue lined by hyperplastic synoviocytes overlying fibro collagenous, fibro adipose tissue

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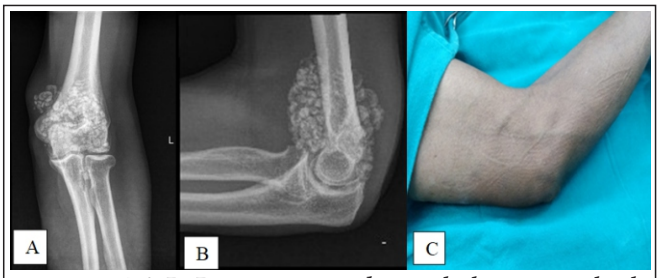

Figure 1: A & B- Preoperative radiograph showing multiple stippled calcifications; C- Showing preoperative range of motion of elbow

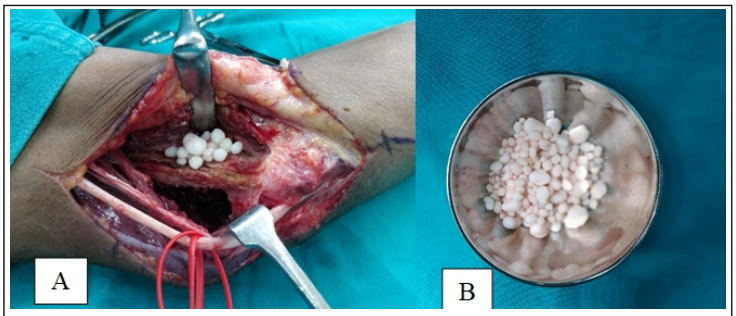

Figure 2: A- Intraoperative picture showing the profile of tumor. B- Post excision with the Bowl full of tumors.

with adjoining fibromuscular tissue showing proliferating capillaries and myxoid change. The section of loose bodies shows fragments of fibrocartilage with chondrocytes surrounded by focal are of ossification, which favored synovial chondromatosis.

At three months, the patient had attained a full range of motion.

Discussion

Synovial chondromatosis is a benign, idiopathic disorder characterized by the involvement of synovial membrane with secondary formation of osteochondral nodules [1]. Following the formation of these nodules in the synovium, fragmentation occurs, resulting in multiple loose bodies formation which are characteristically intraarticular in nature of varying sizes which subsequently gets calcified. Males are commonly involved (57%), with the involvement being classically monoarticular and presentation age ranges between 20 and 50 years. Milgram has described three phases of chondromatosis formation [9]: the active intra-synovial phase with no free bodies formation, transitional phase where there is active intrasynovial proliferation and intraarticular phase where we find numerous loose bodies. As described earlier the monoarticular involvement is classically seen in large joints. Involvement of adolescent and elderly groups is rare. The elbow joint is known to be rarely involved. Mueller et al. reported 20 cases of elbow involvement whereas Khamenei et al reported only 12 cases of elbow involvement respectively [3, 4], and involvement of patients above 50 years was reported only in occasion. In this two large series of 231 cases, lower limb was involved in 70.9% of cases, highest being the Knee (42%) followed by Hip (21.2%) joints. Foot (5.1%), ankle (2.5%), hand (13.8%), shoulder (5.6%), wrist (3.4%) and elbow are less frequently involved [1, 2]. There are only 47 cases reported involving the elbow joints, commonly being unilateral, but bilateral involvement has also been seen [5,6].

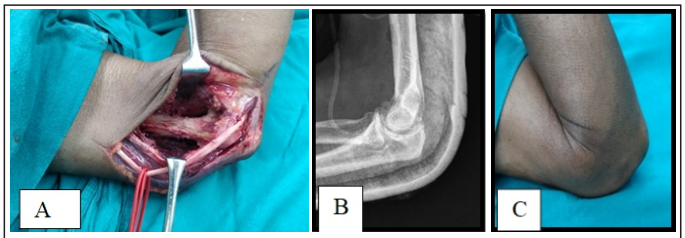

Figure 3: A- Intraoperative picture post excision of tumor. B- Post excision radiograph of elbow. C- Improved range of motion of elbow post operatively.

Various previous studies have mentioned about HLA-DR and CD68 gene expression which directs towards reactive etiopathogenesis of SC. Non-diploid karyo-types, rearrangements, losses, or gains of chromosomes are the other genetic abberations mentioned [9, 10]. However, no cytogenetic analysis was performed in our patient. Patients presents with pain, mechanical symptomss like locking and cluck along with decreased range of movement of joint which are none specific. Pain may follow exertion but may also be due to effusion, locking, nerve compression, or secondary osteoarthritic changes. Stiffness of the elbow is a characteristic of PSC [13], which is one of our patient's symptoms. Nerve compression, especially the ulnar nerve has been described was not present in our patient. These nonspecific symptoms and signs, pose diagnostic challenge to clinician who has to distinguish from other diseases, such as synovial chondrosarcoma, elbow tuberculosis, pigmented villonodular synovitis (PVNS), calcifying aponeurotic fibroma, osteochondritis dissecans, hydroxyapatite deposition, and rheumatoid arthritis [7,8].

Plain radiographs is the investigation of choice which show multiple intra articular smooth rounded loose bodies which are described as "ring and arc" due chondroid mineralisation. subchondral erosion or proliferative arthritis are associated non specific features [13]. CT is more sensitive than X-ray in detecting the ongoing calcification in and around the loose bodies. MRI is mainly useful for identifying the adjacent soft involvement, bone edema and neurovascular involvement. It helps the clinician in deciding between conservative or surgical therapy. Synovial chondromatosis lies in the spectrum between the enchondroma and chondrosarcoma which may explain the rare possibility of recurrences and malignant transformation [1, 7]. Though being described as l locally aggressive with a tendency to recur, it has no metastatic potential. The rate of recurrence is between 7.1% and 15% as described in the

literature [6, 9, 11]. As per reports malignant transformation occurs in only 0.6 percent of cases [2]. The management of SC included both nonoperative and operative modalities. Conservative management has been described in phase one with non-steroidal anti-inflammatory drugs as a part of pain management. However Phase II and III need surgical management [1, 10]. Complete loose body removal is the key for prevention of recurrence which perhaps delay the secondary osteoarthritis [1]. Arthroscopic and open approaches has been describes as a part of operative management. Although various surgical approaches has been described, complete removal of all loose bodies combined with partial or complete synovectomy decides the prognosis [13, 14, 15]. According to Flury et al. [12] both arthroscopic and open techniques provide similar results, but the arthroscopic approach has advantage of shorter rehabilitation period and higher patient satisfaction In preoperative planning, we preferred to consider an open surgical approach rather than an arthroscopic because extensive involvement of articular part of elbow joint and open surgery allows retrieval of multiple large loose bodies and thorough synovectomy can give excellent functional outcome and reducing recurrence chance. Our patient had attained a full range of motion without three months post-surgery.

Conclusion

Primary synovial chondromatosis is a rare and benign condition of the elbow. Symptoms of pain, restriction in elbow extension, and episodes of locking should alarm towards PSC, and appropriate imaging with radiographs and MRI scans should be obtained. Complete removal of loose bodies remains the gold standard as described earlier. Even with extensive involvement, the only symptoms patient presented were related to elbow locking and pain during movement. There were no other symptoms concerning nerve palsy or bursitis in our patient. Surgical synovectomy and complete removal of loose bodies as seen in post operative X-ray have helped to improve the range of motion and function of the elbow joint.

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Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the Journal. The patient understands that his name and initials will not be published, and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

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