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 the formation of multiple cartilaginous bodies. The disease is characteristically monarticular, most commonly involving the knee. Despite frequent mention of elbow involvement in various texts and articles, we wish to review the clinical, radiologic, and histologic features of synovial osteochondromatosis of the elbow 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. The metaplastic process of synovium converts it into the cartilage and gets detached to become a loose body. It mainly affects large joints; knee, hip, shoulder, ankle, elbow, and wrist. Smaller joints have also been involved, including distal radioulnar, tibiofibular, metacarpophalangeal, and metatarsophalangeal joint [1]. Bursae around the joints are also rare critical locations for synovial chondromatosis. It typically presents in the third to fifth decade of life. Patients usually present with pain, swelling, and restriction of movements. Management is mainly surgical, either open or arthroscopic. We present a case of primary synovial chondromatosis in an elderly individual, which is a rare synovial pathology in the elbow.

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.

Based on the clinico-radiological finding, open surgery was planned. Through the Medial approach to the elbow done (Hotchkiss) multiple loose bodies 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 with adjoining fibromuscular tissue showing proliferating

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

capillaries and myxoid change. The section of loose bodies shows fragments of fibrocartilage with chondrocytes surrounded by focal area 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, metaplastic disorder of the synovial membrane with secondary formation of osteochondral nodules [1]. Following the nodules of hyaline cartilage formation in the synovium, fragmentation may occur, resulting in multiple intraarticular loose bodies of various sizes and calcification. PSC has a male preponderance of 57%, typically monoarticular, and presentation age ranges between 20 and 50 years. According to Milgram [9], PSC has been postulated to appear in 3 separate phases: the active intra-synovial phase without free bodies and a transitional phase involving active intrasynovial proliferation and intraarticular loose bodies and multiple loose bodies phase without obvious intrasynovial disease. SC is generally associated with monoarticular involvement, seen in large joints. Involvement of adolescent and elderly groups is rare. It is infrequent for it to involve the elbow joint. In a study by Mueller et al. and Khamenei et al., only 20 cases and 12 cases of elbow involvement were reported in recent years, respectively [3, 4], and involvement of patients above 50 years was only 1. In this two large series of 231 cases, the percentage of involvement was described as follows, the lower limb being affected in 70.9% of cases, primarily the Knee (42%) and Hip (21.2%) joints. Less frequently affected joints are: the foot (5.1%), ankle (2.5%),

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

hand (13.8%), shoulder (5.6%), wrist (3.4%) and elbow [1, 2]. PSC of the elbow is extremely rare, with only 47 cases reported over the years, constituting about 5.2% of reported presentation is mostly unilateral, but bilateral involvement has also been seen [5, 6].

The HLA-DR and CD68 gene expression seen in previous studies supports the view that a reactive condition may play a role in the etiopathogenesis of SC; also many other abnormalities have been reported as non-diploid karyo- types, rearrangements, losses, or gains of chromosomes [9, 10]. However, no cytogenetic analysis was performed in our patient. The clinical presentation is common of pain, mechanical symptoms, and loss of movement, which is 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, damages the sensory and motor of distal limbs which was not present in our patient. Because of the nonspecific symptoms and signs, it's more challenging for clinician to distinguish from other diseases, such as synovial chondrosarcoma, pigmented villonodular synovitis (PVNS), osteochondritis dissecans, calcifying aponeurotic fibroma, elbow tuberculosis, hydroxyapatite deposition, and rheumatoid arthritis [7, 8].

Diagnosis can be achieved with plain radiographs showing multiple intra articular smooth rounded loose bodies with characteristic "ring and arc" chondroid mineralisation and adjacent bones often

have subchondral erosion or proliferative arthritis [13]. CT owns its higher sensitivity than X-ray for the detection of ongoing calcification around or within loose bodies. MRI can accurately reflect the pathological changes of synovial chondromatosis, clear the lesion's scope, and identify the adjacent soft tissue, bone marrow, and neurovascular involvement. It provides invaluable information to the clinician regarding the need of conservative or surgical therapy in those patients suffering from diseases.

Synovial chondromatosis appears to occupy a position between enchondroma and chondrosarcoma which may explain the

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

incidence of recurrences and malignant transformation of this condition [1, 7]. PSC is indeed usually benign, and although it can be locally aggressive with a tendency to recur, it has no metastatic potential. The reported rate of recurrence is between 7.1% and 15% [6, 9, 11]. Malignant transformation rarely occurs as suggested because only 0.6% of chondrosarcomas are secondary to PSC [2]. Both operative and non-operative treatment have been described for the management of SC. Relief of pain has been reported with conservative management with non-steroidal anti-inflammatory drugs in phase I. However, surgical treatment is more advocated in phase II and III [1, 10]. And loose body removal should be done for prevention of recurrence and delay secondary osteoarthritis [1]. Operative treatment has both arthroscopic and open approaches. Irrespective of the surgical approach, removing all loose bodies combined with partial or complete synovectomy is mandatory [13, 14, 15]. Flury et al. [12] reported that both arthroscopic and open techniques could give a satisfactory result, but the arthroscopic approach has many advantages, including a 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. Patients presenting with typical symptoms of pain, restricted extension, and episodes of locking should raise the suspicion of PSC, and appropriate imaging with radiographs and MRI scans should be obtained. Removal of loose bodies and biopsies remains the treatment of choice. 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. Surgical synovectomy and removal of loose bodies 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.

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