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Case report

A RARE SHOULDER DISEASE OF PRIMARY SYNOVIAL CHONDROMATOSIS

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ABSTRACT

Primary synovial chondromatosis is a rare, benign condition which affects the synovial membranes. It commonly involves knee, elbow and hip in young adults but is rare in shoulder. We reviewed a rare case of synovial chondromatosis of the shoulder. A young female came to our hospital with pain and swelling at right shoulder joint. Radiograph, ultrasound and MRI with contrast were obtained. It helped to make a diagnosis of synovial chondromatosis. The shoulder was operated and multiple oval loose bodies measuring up to 8 mm were removed. Partial synovectomy was done.

Keywords: Synovial chondromatosis, Shoulder

INTRODUCTION

Primary synovial chondromatosis affecting the shoulder joint is a rare orthopaedic problem of no known aetiology. It is characterized by the presence of multiple cartilaginous nodules within the joint cavity or are attached to the synovium¹. Typically it affects one of the bilateral joints. Knee joint is the most commonly affected one.^{2,3} Other joints such as the shoulder, elbow, hip, ankle and temporomandibular joints are also affected^{4,5}. Males are the more to be affected predominantly ranging from the third to fifth decade^{1,6,7}. A pre-existing primitive embryonic rest has been postulated to be the etiological basis of the disease². In cultures of fourteen surgical specimens obtained by Jeffrey's in 1967, no growth of

organism occurred. Trauma has been implicated as the precipitating factor for the development of this disease. Neoplastic etiology is considered to be the cause of the cartilaginous metaplasia by some. However a few isolated cases of malignancy have been reported. We here describe an unusual presentation of synovial chondromatosis in the shoulder joint.

CASE REPORT

A twenty four years old female came to orthopaedic OPD with pain in right shoulder. She had pain for 6 months which had precipitated with history of fall. No investigations were obtained after the fall. The pain gradually increased over 6 months. She also had swelling over the shoulder

with restricted movements for 2 months. Radiographs for shoulder were normal. No fracture or dislocation was seen on the radiograph. Ultrasonography study showed moderate fluid at the shoulder joint with multiple well-defined echogenic rounded lesions measuring up to 8 mm within the effusion. There was thickening of the synovium noted. The rounded nodules and thickened synovium showed increased vascularity on Doppler study. The rotator cuff appeared normal

with no tear. MRI with contrast revealed moderate shoulder joint effusion with thickened synovium and multiple enhancing nodules of varying sizes in the joint. The nodules appeared hypointense on T1W and T2W MRI images. Moderate homogenous contrast enhancement of the thickened synovium and nodules was seen. No erosion of the articular surfaces was seen. The rotator cuff appeared intact. Bones showed normal signals.

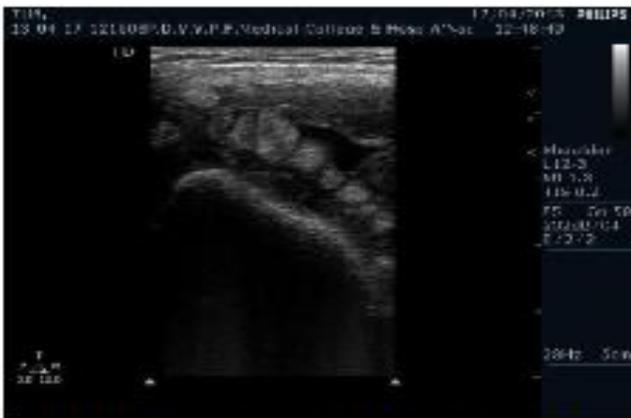


Fig.1: High frequency ultrasound of Rt shoulder shows multiple echogenic nodules, moderate joint effusion.

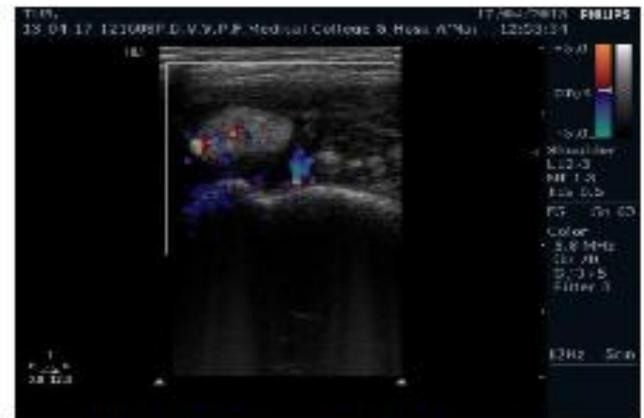


Fig 2: Doppler study of right shoulder shows echogenic nodule with increased vascularity.

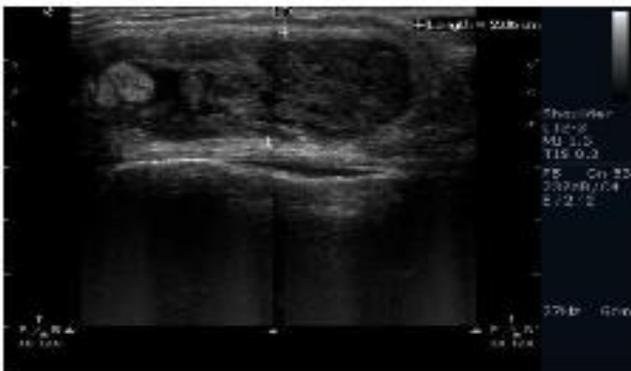


Fig3: High frequency ultrasound of Rt shoulder shows thickened synovium and multiple echogenic nodules.

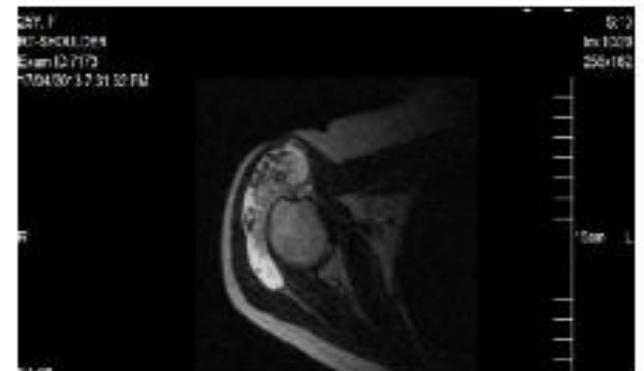


Fig4: T2W axial MRI image shows multiple hypointense nodules with moderate joint effusion

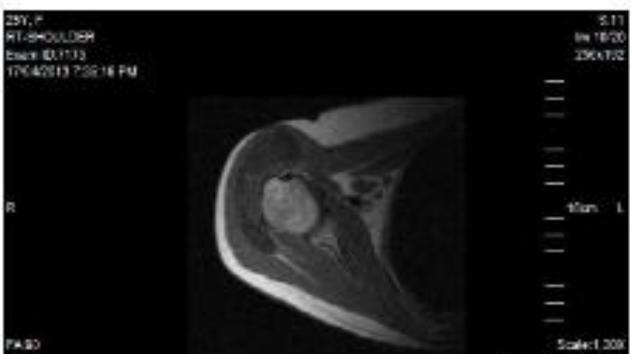


Fig 5: T1W axial MRI image shows multiple hypointense nodules with moderate joint effusion. The articular margins appear regular.

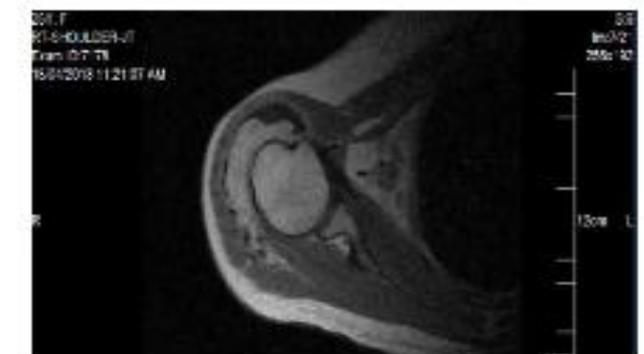


Fig.6: Contrast enhanced T1W axial MRI image shows enhanced hypertrophied synovium and nodules.

DISCUSSION

Synovial chondromatosis is a rare benign condition affecting large joints. Cartilaginous nodules are seen in the synovium of joints, tendon sheaths, and bursae. No history of trauma or infection is noted in majority of the cases. As the disease progresses, the loose bodies may ossify and can be identified radiographically as rounded dense lesions⁸⁻¹¹. Males are affected more than females by synovial chondromatosis. Out of three affected individuals two are males and one is female^{2,11}. It is usually identified in the third to fifth decades of life^{6,7}. The synovial chondromatosis can be differentiated into a primary and secondary form. The primary form occurs in a normal joint unaffected by any other disease⁴. It is characterised by undifferentiated stem cell proliferation in the stratum synoviale. Metaplasia of the synovial cells is considered to be the pathological process. Trauma is commonly thought of as an inciting stimulus however no statistical relationship has been reported in the literature. It has been concluded via immunostaining that primary synovial chondromatosis has a metaplastic etiology¹². The nodules attached to the synovium may get detached as the disease progresses. These form loose bodies in the joint. These loose bodies may continue to grow which are nourished by the synovial fluid. Some of these nodules may calcify and it is called as osteochondromatosis. Secondary condition is said when a joint affected by some other disease also shows changes of synovial chondromatosis. It is thought to be caused by irritation of the synovial tissue¹³. The cartilage fragments get detached from articular surface and become embedded in the synovium to form nodules.

Malignant degeneration is worrying and this has produced recent interest in the diagnosis. Although the change to malignant condition is rare, the patients diagnosed should be monitored³. In a 1998 study examining primary synovial chondromatosis, a relative risk of 5% for malignant degeneration was reported¹⁴. The diagnosis of malignant change

{synovial chondrosarcoma} is made when there is a large, lobulated synovial lesion with extra-articular invasion on imaging studies like MRI or ultrasound. The histological sections show numerous multinucleated cartilage cells having bizarre hyperchromatic nuclei.

Acute primary synovial chondromatosis can be cured adequately by removing the loose bodies and the affected synovium.

CONCLUSION

The shoulder joint of a female is one of the rare joints to be affected and hence this case is reported. Histological diagnosis plays an important role in finding out the malignant change if radiographic or MR imaging are inconclusive.

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