CASE REPORT

A case of idiopathic bursal synovial chondromatosis resembling rheumatoid arthritis

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Abstract

Primary synovial osteochondromatosis is an unusual condition, which generally involves otherwise normal joints. Joints commonly affected in descending order of frequency are knee, hip, glenohumeral joint, elbow and ankle, though any articulation may be involved. Synovial osteochondromatosis has been also encountered in tendon sheaths and periarticular bursa. We report a case with the clinical findings, radiographic features, surgical and histological data of primary subacromial-subdeltoid bursa synovitis. X-ray radiographs of the right glenohumeral joint as well as CT and MRI of the right shoulder zone were performed. A soft tissue mass around the lateral margin of the proximal humerus without evidence of any calcification/ ossification or erosion of the adjacent cortex was detected on both X-Rays and CT images. Multiple nodules of almost equal size appeared that were isointense on T1-weighted spin-echo images and slightly hyperintense on T2 weighted spin-echo images compared with the signal intensity of the surrounding skeletal muscles.

The main differential diagnosis was pigmented villonodular synovitis, rheumatoid arthritis with rice bodies and secondary synovial osteochondromatosis. Based on the results of all modalities the diagnosis of primary synovial chondromatosis of subdeltoid/ subacromial bursa was concluded. Hippokratia 2009; 13 (1): 61-63

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Primary synovial osteochondromatosis is an unusual condition, which generally involves otherwise normal joints. The clinical onset of primary synovial osteochondromatosis varies from childhood to the 7th or 8th decade of life[1]. However it afflicts most frequently men in their 3rd to 5th decade of life. Joints commonly affected in descending order of frequency are knee, hip, glenohumeral joint, elbow and ankle, though any articulation may be involved[1]. The glenohumeral joint is the 3rd most frequent site of involvement[1]. Various patterns of primary synovial osteochondromatosis around the glenohumeral joint have been reported. Occasionally isolated disease of the subdeltoid/subacromial bursa has been encountered[1].

We report a case with the clinical findings and radiographic features data of primary subacromial-subdeltoid bursa chondromatosis.

Case Presentation

A 55 year old male presented at Hippokratio Hospital in April 2006 with a palpable mass on his right shoulder which was first noticed 1.5 year ago. There was no history of trauma or any other inflammatory or infectious disease. On examination there was a mass in the region of the glenohumeral joint but there was no profound restriction of glenohumeral joint motion or tenderness of the palpable mass. No neurological deficits of his right upper arm were found.

Serologic tests for rheumatoid arthritis, other seronegative arthritis or tuberculosis were negative.

Radiographs of the right glenohumeral joint showed a soft tissue mass around the lateral margin of the proximal humerous without evidence of any calcification/ ossification or erosion of the adjacent cortex.

CT imaging the right shoulder zone demonstrated no distension of the shoulder joint or any foci of increased attenuation (calcified areas). Bone erosions, even subtle, were not apparent. CT revealed a soft tissue mass in the deltoid region (Figure 1).

On MR images (1 T scanner) of the same area a markedly distended subacromial-subdeltoid bursa, filled with multiple nodules almost equal size, was found. These nodules were isointense on T1- weighted spin-echo images and slightly hyperintense on T2 weighted fat suppressed spin-echo images compared with the signal intensity of the surrounding skeletal muscles (Figure 2a).On T2- weighted images a small amount of bursal fluid was present. The glenohumeral articulation appeared normal without any signs of degenerative osteoarthritis. The ipsilateral rotator cuff was intact. On Flash 2D gradient echo image no foci of low signal intensity were seen within the markedly distended subacromial-subdeltoid bursa (Figure 2b). According to CT and MRI findings a diagnosis of idiopathic bursal synovial chondromatosis was made.

At surgery, multiple hard shiny bodies were found in the subacromial-subdeltoid bursa. Communication with the ipsilateral glenohumeral joint or obvious damage of the rotator cuff was not observed.

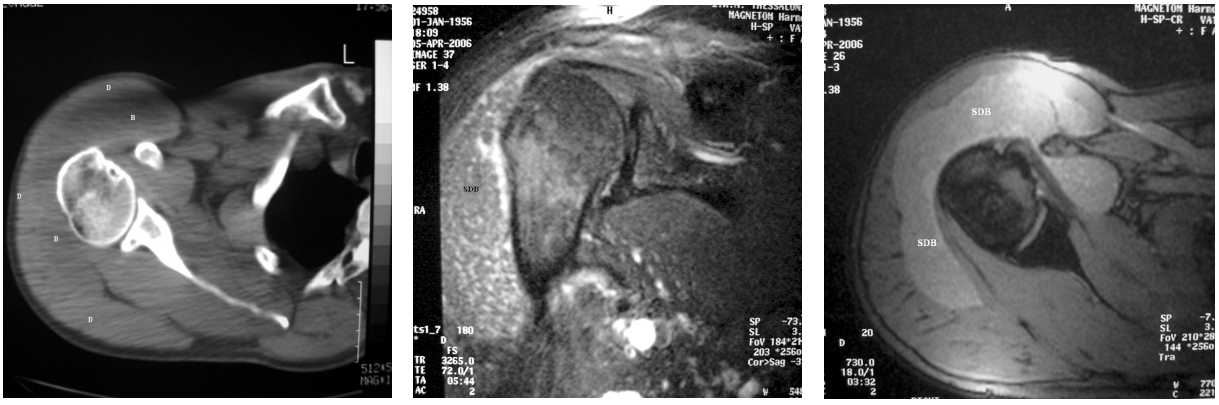

Figure 1: Axial CT image of the right shoulder demonstrates a soft tissue mass in the deltoid region. No calcification or bone erosion is seen (B: Bursa, D: Deltoid muscle).
Figure 2a: Oblique coronal T2 weighted fat suppressed image (TR:3261 ms, TE:72 ms) demonstrates a markedly distended subacromial-subdeltoid bursa (SDB), filled with multiple nodules almost equal size, slightly hyperintense compared with the signal intensity of the surrounding skeletal muscles.

Histological findings confirmed our radiological diagnosis.

Discussion

Synovial osteochondromatosis is an unusual benign metaplastic disorder of the synovium[2,3] which affects usually young and middle-aged men in the 3rd to 5th decade of life. Monoarticular disease is the rule. Most frequent site of involvement is the knee, followed in descending order of frequency by the hip, shoulder, elbow and ankle[1]. Synovial osteochondromatosis has been also encountered in tendon sheaths and periarticular bursa (extra-articular form). Occasionally the process can be extended beyond the joint into adjacent soft tissue

The clinical manifestations of primary synovial osteochondromatosis (also known as idiopathic synovial osteochondromatosis in contrast to secondary degenerative one) are non specific. They include swelling, palpable mass, pain, tenderness, and limited joint motion[1,4].

Complications of synovial osteochondromatosis have been reported. Secondary degenerative osteoarthritis due to chronic mechanical irritation and bone destruction by loose bodies is the rule[5], though rarely malignant transformation to chondrosarcoma has been mentioned in the literature[2,6].

The disease results from metaplasia of subsynovial connective tissue into cartilage nodules[2-4]. Multiple (even hundreds), cartilaginous, loose bodies are commonly formed. Calcification or ossification of the cartilaginous bodies may subsequently occur. Milgram defined three histological phases of synovial osteochondromatosis[3]: a) active intrasynovial phase, b) transitional lesions phase and c) quiescent (inactive) intrasynovial phase.

Synovial metaplasia occurs only in the first and second phase while free fragments are present in the second and third phase.

Radiographic features of primary synovial chondromatosis are quite variable[1,7]. In up to one third of cases of synovial chondromatosis no calcification is apparent on plain radiographs. When no mineralization occurs, x-rays are either normal, or demonstrate a subtle periarticular soft tissue mass. When mineralization occurs, radiographs reveal radiopaque, round or oval, loose bodies within the joint (and infrequently periarticular). Less radiodense centres due to incomplete calcification/ossification of the free fragments may be seen[7].

Computerized tomography has a higher sensitivity than plain radiography for the detection of calcified foci within a joint and/or in a periarticular bursa. The presence of a soft tissue mass, isodense to water, with calcifications (linear or as bodies), either intra-articular or in an adjacent bursa is highly indicative of synovial chondromatosis. Pressure defects at the articular margins are frequently apparent[1].

The MR imaging findings of primary synovial osteochondromatosis are quite variable[1,8-10]. In cases of unmineralized synovial chondromatosis the signal intensity of the process resembles that of fluid; therefore there is risk of misdiagnosed these findings as joint effusion. However the lobulated appearance of the joint process and its slightly inhomogeneous nature are consistent with non calcified or ossified cartilage nodules. In some cases, like ours, the cartilage nodules have signal intensity characteristics that differ from those of fluid. On T1 or intermediate weighted spin echo MR images non calcified cartilage nodules have signal intensity similar to that of muscle with adjacent fluid of lower signal intensity. On T2- weighted spin-echo MR images these nodules show similar or slightly increased signal intensity while surrounding fluid is of high signal intensity[1]. Both inflammatory arthritis (e.g. rheumatoid arthritis) and granulomatous arthritis (e.g. tuberculosis) can lead to a similar MR appearance due to the presence of intraarticular rice bodies[11-15]. In cases of synovial osteochondromatosis foci

of calcification appear as regions of low signal intensity on all sequences. Gradient echo MR imaging may highlight regions of calcification and ossification. With profound ossification structures with peripheral rim of low signal intensity and central regions of higher signal intensity (representing fat or cartilage) are seen on T1 and T2- weighted spin-echo MR images. After intravenous administration of paramagnetic agent heterogeneous enhancement of the thickened synovium and its septa is seen on post contrast T1 weighted MR images[1].

In our case the final diagnosis of synovial chondromatosis of subacromial-subdeltoid bursa was based on negative serologic tests, anatomic location of the process and the following imaging features: 1) absence of calcified intraarticular/periarticular fragments on plain radiographs and CT imaging (in contrast to phleboliths in synovial hemangioma and broken osteophytes in primary degenerative osteoarthritis), 2) preservation of joint space, evident on all modalities (joint space loss is typical in rheumatoid arthritis, primary degenerative osteoarthritis and advanced tuberculous arthritis), c) normal mineralization of periarticular bones (periarticular osteoporosis is seen in rheumatoid arthritis) and d) absence of foci of low signal intensity on all sequences, especially gradient echo MR images, figure 3b (such regions are typical in pigmented villonodular synovitis due to hemosiderin deposits).

The differential diagnosis of primary synovial osteochondromatosis include pigmented villonodular synovitis, secondary synovial osteochondromatosis, rheumatoid or other seronegative arthritis, septic arthritis including granulomatous infection, synovial hemangioma, synovial chondrosarcoma (intra-articular calcification is seen very infrequently with this tumor)[1,2,6,16] and osteochondroma with adjacent secondary bursal osteochondromatosis (which is a rare phenomenon)[1]. In pigmented villonodular synovitis, calcification and ossification are extremely rare while in primary synovial osteochondromatosis only in up to one third of cases, no calcification is apparent. Further more on gradient echo MR images foci of low signal intensity are always encountered in pigmented villonodular synovitis while in primary synovial osteochondromatosis only calcified regions appear as signals of flow void on all sequences[1]. In secondary synovial osteochondromatosis associated with degenerative osteoarthritis cartilage and bone erosions, joint space loss, osteophytes formation, free intraarticular bodies (commonly fewer than 5 and variable in size) are demonstrated. The affected population is also at least middle aged men and women[5]. In rheumatoid arthritis early joint space narrowing and periarticular osteoporosis are seen. Serologic tests are positive[11,13-15]. Concerning granulomatous infection (e.g. tuberculous arthritis) a prior history of tuberculosis is helpful for the final diagnosis[12]. Both inflammatory arthritis (e.g. rheumatoid arthritis) and granulomatous arthritis (e.g. tuberculosis) can occasionally lead to an MR appearance (intraarticular rice bodies) similar to that of non calcified synovial chondromatosis[13-15]. Subsequently the final diagnosis can not be made exclusively by MR imaging. Finally MR features of synovial hemangioma are diagnostic of this entity even though plain radiographs are not helpful for the final diagnosis of hemangioma, over synovial chondromatosis, due to the similarity between phleboliths and calcified loose bodies[1].

The treatment of synovial chondromatosis is surgical[4,5,17]. Open surgery or arthroscopic intervention is performed with resection of the diseased synovium and removal of any loose intra-articular bodies. Recurrence is frequent after partial synovectomy, hence total synovectomy has been suggested as the preferred treatment.

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