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Reichel Syndrome of Hip: A Rare Entity

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Abstract: Reichel syndrome otherwise called as synovial osteochondromatosis is a rare disease that creates a benign change or proliferation in the synovium or joint-lining tissue, which changes to form bone forming cartilage. Many treatment options like an open synovectomy with removal of the loose bodies, removal of the loose bodies only, a radical synovectomy, an arthroscopic synovectomy with removal of the loose bodies and total hip replacement with radical synovectomy have been reported in various literatures. In this article we are going to discuss about a rare case of Reichel syndrome of hip treated with total hip replacement.
Keywords: Reichel syndrome, Synovial osteochondromatosis, Total hip replacement.

INTRODUCTION

Reichel syndrome otherwise called as synovial osteochondromatosis is a rare disease that creates a benign change or proliferation in the synovium or joint-lining tissue, which changes to form bone-forming cartilage. Primary synovial osteochondromatosis has an incidence of 1:100,000. It is usually a monoarticular disease, although polyarticular involvement is reported in up to 5% of cases [1]. The majority of intraarticular disease involves the knee (50%), with the hip, elbow and shoulder less commonly affected Primary synovial chondromatosis of the hip joint is rare, and the optimal treatment is still controversial. Many treatment options like an open synovectomy with removal of the loose bodies, removal of the loose bodies only, a radical synovectomy, an arthroscopic synovectomy with removal of the loose bodies and total hip replacement with radical synovectomy have been reported in various literatures. In this case we are going to discuss a rare case of primary synovial chondromatosis of hip treated with total hip replacement.

CASE REPORT

A 56 yr old male patient presented with complaints of progressive pain and stiffness of both hips, more on the left than on the right for the past 8 yrs. He was a tailor by profession and has retired due to his painful hip condition. The pain started insidiously and progressed slowly over past eight years. It was dull aching in nature, persistent and aggravated on movements, non-radiating. He has complaints of night pain and was able to sleep only in a sitting posture. Pain started on the right hip and appeared on left hip six and half years back. Now he has more pain and stiffness in his left hip. He is an alcoholic on abstinence for the past 4 yrs. He also has a history of pulmonary tuberculosis treated medically for 6 months 14 yrs back. Radiograph of pelvis with both hip shows severe articular destruction of bilateral femur head and acetabulum (Fig. 1). Our differential diagnosis were avascular necrosis femoral head and tuberculosis hip. Patient underwent total hip replacement after discussing the pros and cons of the surgery with patient and patients relatives. At time of surgery, multiple free and embedded intra-articular chondromatoses were seen and removed (Fig. 2, Fig. 3). Complete synovectomy and debridement was done and specimen were sent for biposy. Histological evaluation showed focal chondroid metaplacia [1-3] and no signs of tuberculosis or active infection, confirming synovial osteochondromatosis. The postoperative period was uneventful. Patient is having good pain free range of movements in left hip (Fig. 4).

DISCUSSION

In Reichel syndrome or Synovial chondromatosis, cartilaginous metaplasia takes place within the synovial membrane of the joint, which organizes into nodules. With minor trauma, nodules are shed as small bodies into the joint space. In some patients, the disease process may involve tendon sheaths and bursal sacs. Cartilaginous intra-articular bodies float freely within the synovial fluid, and derive nutrition from the synovial fluid and continue growth. Progressive enlargement and ossification occur with time. If they remain free, they continue to grow larger and more calcified [1, 2]. In severe cases, they may occupy the entire joint space or penetrate to adjacent tissues. Also,

they can deposit in the synovial lining, reestablish a blood supply, and become replaced by bone. On occasion, synovial reattachment can lead to complete reabsorption of the cartilage fragment. Due to the rare nature of conditions, diagnosis is delayed till mechanical obstruction and damage occurs. As was the case in this patient, who presented late with synovial osteochondromatosis of both hips which occurs in older patients in joints previously affected by joint disease such as osteoarthritis. This pattern of late presentation was seen in several other studies [3, 4].

Controversy exists regarding treatment of synovial chondromatosis, especially with regard to the hip. Localized disease can be treated with resection of affected synovium using arthroscopic synovectomy and debridement [4]. But in our patient with generalized intra-articular disease and severe restriction of motion we did open complete synovectomy followed by total hip arthroplasty due to presence articular damage of femoral head and acetabulum. Total hip arthroplasty here gives the surgeon the chance to address the synovial disease, and also articular damage as well.

Hip arthroscopy can be a useful treatment for synovial chondromatosis. Boyer and Dorfmann [5] reported hip arthroscopy to be curative in 62% of patients at 6 years. However, 38% required additional surgery that includes repeat hip arthroscopy, open synovectomy and debridement, and hip replacement [4]. The authors concluded that as many patient required additional surgeries,complete arthroscopic synovectomy is unlikely [6].

Open synovectomy and removal of loose bodies has been used for treatment. Lim et al [4] reported it to be successful in 17 of 21 patients at 4.4-year follow-up. In order to prevent recurrence surgical dislocation is needed for complete synovectomy [6].

Schoeniger *et al*. [7] reviewed eight patients with monoarticular synovial chondromatosis of the hip having joint débridement and a modified total synovectomy was performed through a surgical hip dislocation with a trochanteric flip osteotomy. Results suggested that open debridement with modified total synovectomy as an effective treatment for preventing recurrence and providing substantial pain relief.

Thomas shitzer *et al*. [8] treated 31 patients with synovial chondromatosis: 12 had synovectomy and removal of loose bodies, and 16 had removal of loose bodies only. 2 had total hip replacement and 1 had resection arthroplasty.

Duncan Ackerman *et al*. [9] reported treatment of 7 patients with total hip arthroplasty and 4 patients with total knee arthroplasty concluding total joint arthroplasty as a valuable treatment option for these patients with predictable improvement in pain and function. Patients remain at risk for recurrence.

Fig. 1: Pre operative X-ray

Fig. 2: Intra operative loose bodies

Fig. 3: Loose bodies eith synovium

Fig. 4: Post operative X-ray

CONCLUSION

Though there are many forms of surgical management of Synovial osteochondromatosis available like arthroscopic partial synovectomy and removal of loose bodies, we opted for total hip arthroplasty. Patient had improvement in range of movements and pain relief on the operated site. He has bilateral pathology and patient is now presurising us for total hip arthroplasty of the opposite side.

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