**Ahlback’s disease: Spontaneous osteonecrosis of the Knee.**

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**ABSTRACT**

Ahlbäck disease is a clinical syndrome characterized by bone necrosis of a load-bearing portion of the femoral condyle, followed by subchondral fracture, subsequent segmental collapse and arthrosis. The diagnosis of the Ahlbäck disease is challenging as sometimes it mimics infection and receives treatment for long time. We report a case of 36 year old gentleman, who presented with confusing radiological picture.

**INTRODUCTION**

It is a clinical syndrome characterized by bone necrosis of a load-bearing portion of the femoral condyle, followed by subchondral fracture, subsequent segmental collapse and arthrosis. This was described by Ahlbäck et al as a distinct clinical entity in 1968¹. The diagnosis of Ahlbäck disease is really challenging and bases on various modality of investigations. We report a case of 36 year old gentleman who has confusing radiological pictures between infection and tumor. The final diagnosis was made by bone biopsy and histopathology.

The aim of this report was to emphasize the need of vigilance required in making a correct diagnosis as many rare conditions also mimic common conditions.

**Case Report**

A 36 year old gentleman, presented with 8 months history of pain over and around left knee. The pain started after a trivial trauma which he sustained about 9 months back while working on field. The pain was mild to moderate, gradually increased in intensity, non-radiating and was aggravated by activities and relieved by rest, pain was worse at night. There was diffuse swelling of knee with supra patellar fullness. The skin overlying the knee joint was normal and there were no scar and sinuses. The skin overlying the knee joint was warmer than the normal side. The lateral femoral epicondyle was tender to deep palpation. There was moderate knee effusion and the knee range of motion was comparable to the normal side however it was painful in its terminal flexion.

A plain x-ray of the left knee joint was done which revealed diffuse irregular thickening of lateral femoral condyle (Figure 1, Arrow). A CT Scan was ordered which showed ill-defined lytic sclerotic lesion in lateral femoral condyle and the suggested a radiologic picture of chronic osteomyelitis (Figure 2).

![Figure 1. X-Ray Left Knee](image-url)

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When he presented to us he was already having Flucloxacilline for last 12 weeks but his symptoms were not getting better. He still had the swelling and there was quadriceps wasting of 1.5 cm. we repeated his blood parameters (Hb%, ESR and CRP) which were all within normal limits. We requested for MRI of the left knee joint which showed focal lesion in the lateral condyle of left femur including the subarticular, epiphyseal as well as the mid diaphyseal areas showing low signal intensity in T1W and heterogenous signal intensity in T2W and hyper signal intensity in STIR sequences and other smaller similar character intramedullary lesion in the mid-distal diaphysis and the lesion appears contiguous in STIR sequence (Figure 3 and 4). The MRI also suggested a possibility of infective pathology, however the mentioned that there were no definitive features of chronic osteomyelitis.

The radiological reports were confusing and considering the lesion has reached its present size in 8 months duration we suspected a tumor lesion and decided to perform open biopsy.
Biopsy was performed through a lateral incision. There was knee effusion, the synovial fluid was collected and sent for PCR to detect tuberculosis. There were no periosteal reaction and pus subperiosteally. And nothing came out during drilling either. A conical shaped bone piece was removed and few pieces of soft tissues from and around the lesion was also taken and sent for histopathological examination.

Synovial fluid examination revealed exudative reaction without any pus cells, PCR was negative for tuberculosis. Soft tissue histology showed only mild hyperplasia of the
synovium. The bone biopsy reported as foci of trabecular and bone marrow necrosis. The necrosis is localized and is surrounded by a band of fibrovascular granulation tissue of variable thickness, with focal areas of osteoclastic activity and histiocyticresorption of the dead tissue. These feature consistent with Ahlbuck's disease. The patient started feeling less pain after the surgery and was treated with analgesics and exercises only.

**DISCUSSION AND LITERATURE REVIEW**

With the histopathological diagnosis of bone infarct and ruling out infection, tuberculosis and any tumor lesion, we started doing literature search. The literature seemed to use various terms like Ahlbäck disease, Spontaneous Osteonecrosis of Knee (SPONK, SONK), Osteonecrosis, Osteochondritis dissecans, secondary osteonecrosis. Ahlbäck et al. first reported on osteonecrosis of the knee in 1968. The osteonecrosis that Ahlbäck described now is referred to as spontaneous osteonecrosis of the knee (SPONK).

There were various theories hypothesized for the etiology of SPONK. The theories include vascular insufficiency or microfractures of the subchondral plate, with deviation of the synovial fluid from the synovial space to inside the bone. The knee is the second most common site for SPONK, but it is affected much less often than the hip. The true incidence of the disease is unknown, but osteonecrosis of the knee is believed to account for approximately 10% of cases of osteonecrosis. SPONK was found to be affecting female over the age of 55 who are obese, however our patient was a male of 36 years and was thin built. The pain worsens at night, limits physical activity and does not improve with physiotherapeutic treatment or use of nonsteroidalanti-inflammatory drugs, which was very similar to our case.

There was diagnostic challenge in our case as the lesion was very big involving even distal femoral diaphysis. The SPONK was said to occur unilaterally in 99% of cases as of our but it was reported to be more common on the medial femoral condyle in contrast to our case where whole of the lateral condyle was involved.

Aglietti devised a classification system for SPONK, and our case fitted to Stage 3 (Table 1). Studies have shown the prognosis of spontaneous osteonecrosis of the knee (SPONK) to be related directly to the size of the lesion. Aglietti et al. reported that lesions greater than 5 cm² had a worse prognosis than lesions with areas less than 3.5 cm². Lotke et al. reported that lesions occupying greater than 50% of the femoral condyle have a worse prognosis. Prognosis has also been shown to be worse in advanced-stage lesions.

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
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<tr>
<td>I</td>
<td>Plain radiograph findings are normal. Diagnosis must be made from MRI or bone scan.</td>
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<tr>
<td>II</td>
<td>Radiographs show flattening of the weightbearing portion of the condyle.</td>
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<tr>
<td>III</td>
<td>Radiographs show a radiolucent area surrounded by sclerosis.</td>
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<tr>
<td>IV</td>
<td>Radiographs show a more defined ring of sclerosis and subchondral bone collapse forming a calcified plate, sequestrum, or fragment.</td>
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<td>V</td>
<td>Narrowing of the joint space, osteophyte formation, and/or femoral and tibial subchondral sclerosis is shown.</td>
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Treatment of SPONK still remains controversial and said to be guided by the stage. Surgical options include Core Decompression, High tibial osteotomy, and Knee Arthroplasty. If there are joint destruction probably an arthroplasty is choice of treatment. But if the joint architecture are maintained then core decompression may be the treatment. The principal behind core decompression is reduction of interosseous pressure, thereby restoring adequate circulation. Forst et al. treated five patients with early stage lateral spontaneous osteonecrosis with core decompression and reported it to be an effective treatment. When we did a core biopsy and removed a conical bone piece this acted as a core decompression for the patient which may be the reason why patient improved clinically.
REFERENCES


