# Systemic Mastocytosis Diagnosed Following Bone Biopsy During Total Knee Replacement: A Case Report

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**Citation**

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**Abstract**

Mastocytosis is a rare, heterogeneous disorder, of normal, active mast cells. The increase in mast cells is usually localised to the skin, but occasionally occurs in other organs as a systemic disorder. We report a case where systemic mastocytosis was diagnosed following biopsy of the femur during total knee replacement.

## CASE REPORT

A 60 year old postman presented in Jan 2002 with a two-year history of a painful swollen left knee. His walking distance was a quarter of a mile and he had night pain. He had no medical history of note and was taking non steroidal anti inflammatory medication for pain relief.

On examination he looked well in himself. He had a cutaneous macular rash involving his neck; trunk and limbs .He had a palpable spleen, 2-3cms below the left costal margin. His left knee was swollen and he had medial joint line tenderness and the Mc Murray's test was positive. The range of movements was from 10-90 degrees of flexion with a fixed flexion deformity. X-Ray's of his left knee showed osteoarthritis.

In Feb 2003 he underwent a left total knee replacement. Intra-operatively the synovium around the knee was found to be inflamed. It was initially not possible to pass the intramedullary rod into the femur despite normal looking x-rays of the femur. Also the femoral cancellous bone appeared to be dark brown. On the basis of these findings the femur was biopsied and the procedure was completed using the limited portion of intramedullary rod available for alignment. He was slow to mobilise postoperatively but made an uneventful recovery and he was discharged on the 17th postoperative day with a range of movements from 0-90 degrees.

The histopathology report of the bone biopsy showed that in the marrow of the subchondral bone there was evidence of Systemic Mastocytosis. In addition a Bone marrow trephine biopsy from right posterior iliac crest confirmed Systemic Mastocytosis. He also had a CT of his abdomen which showed a moderately enlarged spleen and an assessment for leukaemia showed apparently normal male karyotype. Blood investigations were normal and he is being followed up regularly by the oncologists. He continues to be well in himself 22 months after his surgery.

## DISCUSSION

The presence of too many mast cells or mastocytosis can occur in 2 forms, cutaneous and systemic. It has been reported to be present in one of 1000 to 8000 new patients attending dermatology clinics ([1,2]).

The disease may be benign, with minor transient symptoms and signs that never cause the patient to consult a physician, or it may be life threatening. In our case the patient had a macular rash for approximately 40 years, which never caused him any discomfort and did not need medical attention.

Approximately 90% of patients with mastectomies manifest only cutaneous lesions and do not have serious haematological disorders. Cutaneous mastocytosis has a low incidence of systemic involvement in children, whereas systemic mastocytosis occurs in more than 25 % of adults ([3,4]).

Many patients with mastocytosis have a long history of chronic symptoms that are occasionally punctuated by acute exacerbations ([5]). In one series of adult patients the average interval between onset of symptoms and the correct diagnosis was 10 years ([6]).

In our report the patient gave a history of occasional bone pains and long-standing cutaneous rash, which was not pruritic but became worse during hot weather.

Laboratory abnormalities associated with systemic mastocytosis include hypocholesterolemia (10% - 20 % of the patients), thrombocytopenia, eosinophilia (12%-25% of patients), increase in alkaline phosphatase, and elevated liver enzymes may also be present in patients with systemic disease. Circulating biochemical mediators are often increased especially when a large number of mast cells are activated during attacks ([7]). Although several biochemical tests of blood and urine have been used to support the diagnosis of mastocytosis, one of the most sensitive when it is available is an elevated plasma tryptase level ([8]). The patient in our case report had almost all his blood investigations within normal range.

## CONCLUSION

Knee joint replacement is a common operation. Though comment is occasionally made relating to the quantity and the quality of bone and cartilage, the main concern is to ensure good surfaces and cuts for the application of the prosthesis.

Bone cysts are often filled with cement and synovium is quickly reflected or resected. Odd appearances are attributed to the deforming nature of osteoarthritis and similar joint specific inflammation. This case highlights the importance of tissue diagnosis if there doubt regarding the nature of bone or other tissues encountered in joint replacement.

To our knowledge this is the only case to be reported where Systemic Mastoctosis was diagnosed following biopsy of the femur during total knee replacement. Interestingly, despite normal x-rays of the femur, we were unable to pass an intramedullary rod down the femur. It was passed one third of the way down and despite considerable force we were unable to pass it any further. The biopsy was conducted as a result and led to the diagnosis of systemic mastocytosis. Therefore, is there an indication to biopsy bone during a total knee replacement if it is not possible to pass an intramedullary rod for no apparent reason? This case report would suggest there is an indication to do this.

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