

Total Hip Arthroplasty in a Patient with Systemic Mastocytosis

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INTRODUCTION

Mastocytosis is an uncommon myeloid malignancy involving mast cells (MC) and their CD34+ progenitors. Signs and symptoms are caused by the release of biological mediators from MC and infiltration of neoplastic MC in different organs. The skin and the bone marrow are predominantly involved.

Total Hip Replacement (THR) in a patient with systemic mastocytosis (SM) is a rare occurrence. We present a case of a 38-year-old caucasian male with systemic mastocytosis who underwent THR.

CASE REPORT

A 38-year-old male with SM was referred to our outpatient clinic with severe pain in the left hip, especially on weight-bearing. Systemic mastocytosis was diagnosed 18 years previously. He had urticaria pigmentosa skin lesions (Fig.1) and signs of bone marrow infiltration including bruising, bleeding and anemia. Physical examination revealed an antalgic gait and a decreased range of motion without flexion deformity [flexion - 90°; abduction - 15°; adduction, internal rotation and external rotation - 10°]. There was no limb length discrepancy. The Harris Hip Score [HHS] was 48 of 100 [poor].

Anteroposterior pelvic radiograph showed diffuse sclerosis and the medullary canal seem obliterated. Obvious features of osteoarthritis were present in the left hip with loss of the joint space and periarticular cyst formation were present.

Interestingly there were no osteophytes (Fig.2). A CT scan of the proximal femur demonstrated an extremely narrow medullary canal (Fig.3). The MRI excluded avascular necrosis.

The general condition of the patient was good and the life expectancy was considered reasonable, therefore a cemented THR was performed [posterior approach]. We observed significant bleeding from the soft tissues. The femoral neck osteotomy was done without any trouble, but the definition of the medullary canal was difficult (Fig.4), whereby the preparation of the femur was done using fluoroscopy. The implantation of the acetabular component ran smoothly.

Histological examination (Fig. 5 and 6) of the marrow component showed an increase in the cellularity with multifocal, sharply demarcated, compact infiltrates of MCs consistent with the disease. The patient had an uneventful postoperative course. With three years of follow-up, he has a normal gait; slight, occasional pain, with no compromise in activity; HHS 96 of 100 [excellent]. There are no radiological signs of loosening.



Fig.1 – Urticaria pigmentosa



Fig.2 – Radiograph of the pelvis –sclerotic bone. Degenerative disease of the left hip with loss of joint space and cyst formation.

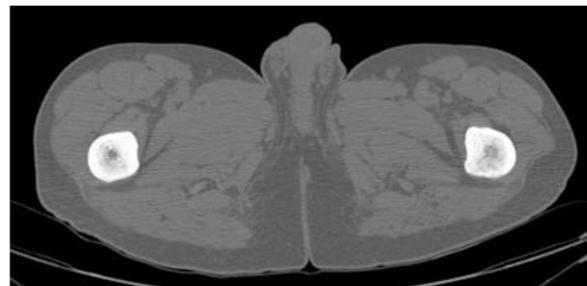


Fig.3 – CT scan below the level of the lesser trochanter – uncommon thick cortices with a narrow medullary canal.



Fig.4 – Cut surface of the femoral neck – amorphous material without evidence of the canal

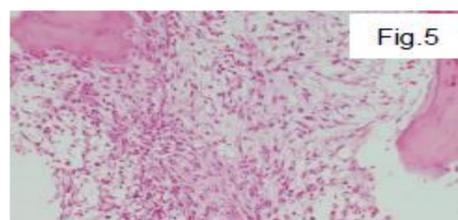


Fig.5 – Systemic Mastocytosis (HE,x100) The marrow compartment showing diffuse infiltration with abundant polygonal / spindle mast cell.

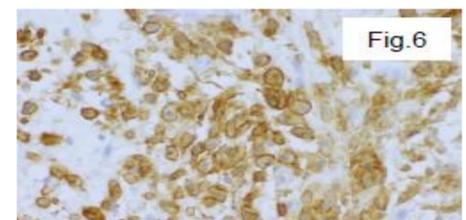


Fig.6 – Systemic Mastocytosis (CD117,X 400) Mast cells with strong expression of CD117 (c-Kit)-membrane immuno-reactivity.

DISCUSSION/CONCLUSION

Before surgery, our main concern was to decide the appropriate method of fixation. We opted for a cemented fixation after reviewing the experience of other authors. To date, there are only **two cases reported in the literature of patients with SM undergoing THR**. According to this results and having in mind that osteoporosis is the most prevalent bone manifestation in this scenario, we considered the potential for osseointegration low. Intraoperative, the **recognition of the medullary canal** was difficult due to the unusual bone density. Intraoperative fluoroscopy was valuable to get the correct alignment and size of the femoral component.

Although particular concerns will persist regarding the pathophysiology of SM, the followed strategy provided good clinical and radiological results at three years.

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