Systemic mastocytosis is a rare condition that often involves the bone marrow. We report the case of a patient with systemic mastocytosis who underwent total hip replacement. Technical difficulties encountered during the procedure included a narrow medullary canal and abnormally hard bone, later confirmed by laboratory measurements. Follow-up at five years showed a good clinical and radiological outcome.

Systemic mastocytosis is a condition characterised by mast cell hyperplasia or neoplasia, and its true incidence is unknown. Cutaneous manifestations such as urticaria pigmentosa may be present. In most cases with systemic disease the bone marrow is usually involved. The long bones are more commonly affected and may appear both sclerotic and demineralised on radiographs. Skeletal changes are present in 70% of patients with systemic disease, and may be diffuse or circumscribed. Typical radiological appearances are of stippled, mottled or granular bone texture. Osteoarthritis can also coexist with the condition.

Mast cell degranulation and the release of mediators, especially histamine and prostaglandin D\(_2\), can be precipitated by triggers such as trauma, extremes of temperature and drugs, including morphine, codeine, acetylsalicylic acid and alcohol. This can lead to profound cardiovascular collapse and bronchospasm. The peri-operative management and anaesthesia can be challenging, and the pre-operative use of antihistamines and sodium cromoglycate has been advocated.

Hip replacement has been described in mastocytosis, but with a poor outcome at one year. We describe a case of systemic mastocytosis with associated osteoarthritis of the hip treated with a cemented total hip replacement (THR). There were concerns pre-operatively that the bone would be too dense and hard to allow suitable preparation for the implantation of a prosthesis. However, this proved not to be the case, and so an investigation into the mechanical properties, histology and histomorphometry of the bone was undertaken.

In view of the lack of knowledge concerning the mechanical properties of mastocytosis of bone, measurements of hardness were performed on the retrieved femoral head and compared with a standard arthritic femoral head.

**Case report**

A 64-year-old woman presented with pain in the right hip and a coexistent diagnosis of systemic mastocytosis. The diagnosis had been made 12 years previously. She had anaemia and occasional skin rashes. Her pain was resistant to conservative measures and she was referred with a view to THR. There were no other associated joint problems. She walked with the aid of a stick and had severe pain on weight-bearing. She had an antalgic gait. Examination of the right hip showed a fixed flexion deformity of 20°, with flexion to 90° and gross limitation of rotation. The Oxford Hip Score was 49 of 60, with 12 of 60 being normal function and 60 of 60 being severe disability. Plain radiographs showed sclerotic bone with apparent obliteration of the marrow cavity. There was loss of joint space and periarticular cyst formation consistent with osteoarthritis of the hip (Fig. 1). There was concern that the apparent lack of a medullary canal would prevent the use of a stemmed femoral component. A CT scan of the proximal femur demonstrated a potential medullary canal, although the diameter was small (Fig. 2).

A resurfacing-type arthroplasty was considered as an option, but a conventional stemmed femoral component was thought to be the best solution for achieving long-term fixation in an abnormal bone.

No information concerning the hardness of bone in systemic mastocytosis was available,
and the radiographic appearances also suggested the possibility of difficulty during preparation of the acetabulum.

During the operation there was noticeable bleeding from the soft tissues, but haemostasis was achieved with care. The hip was dislocated via an anterolateral approach and the femoral neck divided with an oscillating saw. Although the bone felt harder than normal on resection, no significant difficulties were encountered. The acetabular cartilage was denuded and preparation of the underlying subchondral bone was only possible with a high-speed burr, rather than with standard acetabular reamers. Pits and troughs were created to allow the cement to intrude. Cement was used for fixation, because the potential for osseointegration with an uncemented component was unknown, bearing in mind the underlying pathology. There was minimal bleeding from the surfaces of the prepared bone, and a standard Charnley polyethylene acetabular component (DePuy, Leeds, United Kingdom) was introduced with Palacos R cement (Schering-Plough Ltd., Welwyn Garden City, United Kingdom) and appropriate pressurisation.

Definition of the medullary canal of the femur was difficult. The appearance of the cut surface of the femoral neck was one of amorphous bony material without evidence of a canal. With great care, a 5 mm diameter high-speed burr was used to define an opening in the piriform fossa. Once this was achieved, it was possible to identify a narrow medullary canal using ball reamers at a lower speed. Standard Exeter (Stryker Howmedica Ltd, Newbury, United Kingdom) rasps were then used for sizing purposes, but they were ineffective at removing bony material. Further careful use of the ball reamers widened the available canal, and a size zero 37.5 mm offset Exeter femoral component was tested and found appropriate for insertion.

Channels and troughs were fashioned to produce a rough surface for cement intrusion. A distal cement plug was not necessary, and the chosen Exeter component was inserted following lavage and pressurisation of Palacos R cement. The post-operative course was uneventful.

When seen after five years the patient reported complete resolution of the pain in her right hip, although she was developing symptoms from the left hip. She walked without a stick or a limp and was satisfied with the outcome. The Oxford Hip Score at five years was 15. There were no radiological signs of loosening, no bone resorption and no radiolucent lines at the bone-cement interface (Fig. 3).

**Hardness measurements.** Microhardness measures the resistance of a material to being deformed by a small diamond indenter. It is useful for measuring specimens that are small or convoluted and from which standard mechanical testing specimens cannot be fashioned. ‘Hardness’ is not directly related to stiffness or strength, but has been found to have a reasonably close relationship to Young’s modulus (a measure of stiffness) and the yield strength. It is a material rather than a structural property of bone. It might be expected that, if the bone had been very hard, it would have been difficult to cut during surgery.

The femoral head from an uncomplicated hip replacement performed in a 68-year-old patient was used as the control. A sagittal slice roughly 3 mm thick was cut from each femoral head. The slices were then polished with a series of increasingly fine carborundum papers and finished to a glassy polish with 0.05 µm alumina paste.

Microhardness measurements were determined with a Leitz Wetzlar ‘miniload’ hardness tester (060-366.002; Ernst Leitz GMBH, Wetzlar, Germany). An indenting load
of 25 pond (25 g weight) with a descent time of 15 seconds for the diamond was used. Once it reached the lowest position the diamond was allowed to impress the bone surface for a further ten seconds. The specimens were tested dry because it was important to place the indent centrally on the trabeculae, and this would have been difficult if the specimen was wet. However, testing dry is also not ideal, because the greater hardness of dry as opposed to wet bone has been demonstrated previously. However, the difference is not great: Blackburn et al\(^7\) found that in human cancellous bone dry testing gave values of about 20% greater than wet testing.

The product of the length of the two diagonals was converted to a Vickers hardness number using a standard formula.

A total of 20 indents were made on each slice. The results for each were distributed quite tightly and were approximately normal (Table I).

Although the difference between the means was significant it was not large, the hardness of the mastocytotic bone being only about 14% greater than that of the osteoarthritic bone. These values for hardness are very similar to those reported by Blackburn et al\(^7\) for the hardness of normal human trabecular bone (mean Vickers hardness number, 44.9, \(n = 90\)).

**Histology.** Histological examination of the decalcified bone showed surface osteosclerosis and cartilage destruction consistent with osteoarthritic change. There was a profound increase in the cellularity of the marrow compartment, with abundant polygonal/spindle mast cells which showed weak staining of the cytoplasmic granules with Toluidine blue (Fig. 4). There was associated marrow fibrosis, with sparse islands of haemopoietic cells showing all three normal lineages. The bone volume was markedly increased compared to normal\(^8\) (58% of total bone volume; normal 16% \(\pm\) 4%). The morphometry was investigated using Adobe Photoshop software (Adobe Systems Inc., San Jose, California) by assessing pixel histograms of digitised photomicrographs.

<table>
<thead>
<tr>
<th>Bone</th>
<th>Number</th>
<th>Mean VHN</th>
<th>sd</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mastocytosis</td>
<td>20</td>
<td>50.7</td>
<td>5.19</td>
</tr>
<tr>
<td>Osteoarthritis</td>
<td>20</td>
<td>44.6</td>
<td>6.75</td>
</tr>
</tbody>
</table>

\(t_{38}\) value of difference between means = 3.21, \(p = 0.003\)

\* VHN, Vickers hardness number

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**Fig. 3**
Radiograph five years post-operatively showing a cemented hip arthroplasty with no radiological evidence of loosening or subsidence.

**Fig. 4**
Photomicrograph of trabecular bone (Oss) showing an increase in the bone volume. The marrow compartment contains both normal haemopoietic (H) and mastocytosis tissue (M) (Masson trichrome, x 40). The inset (b) shows granular cytoplasmic Toluidine blue staining of mast cells.

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Table I. Results of the Vickers hardness tests\(^7\) on the two slices.
Discussion

Systemic mastocytosis is a rare condition, and there is only one case of a hip arthroplasty associated with it reported in the literature.\(^4\) The case described here gave difficulty in deciding the correct choice of surgical technique, the method of fixation and the prosthesis. In the case mentioned above, an uncemented component failed rapidly within a year.\(^4\) The reason for this was not clearly stated, but may relate to the failure of osseointegration into the uncemented implant.

In the absence of information concerning the biological behaviour of mastocytosic bone in the presence of implanted material, cement was used for fixation. The porosity of the bone seen intra-operatively was poor. It was felt that significant cement intrusion on a microscopic scale would be unlikely. It was possible to provide a macroscopically rough surface, with the use of burrs, onto which the cement might key. This has probably contributed to the success of the procedure in the short term.

There was an initial concern that the apparent density of the affected bone would make sawing and preparation difficult, but this proved not to be the case. In the event, the measured hardness of the trabecular bone was only about 14% greater than that of bone from a control osteoarthritic femoral head (mean Vickers hardness number 50.7 (SD 5.19) vs 44.6 (SD 6.75)), so it is not surprising that little difficulty was encountered on cutting the femoral neck. The hardness of the bony trabeculae in the osteoarthritic femoral head was also similar to that in trabeculae from the ‘normal’ bone.\(^7\) The difficulties that were encountered in creating a channel in the medullary canal had much more to do with the density of the bone, rather than its intrinsic hardness. Nevertheless, great care had to be exercised in the preparation of the medullary canal of the femur, as it was only possible to remove the bone using powered instruments.

The use of a cemented THA in this case provided good results clinically and radiologically at five years. The possible technical problems were not as difficult to overcome as might have been expected. Although the bone was undoubtedly dense (58% of total bone volume compared to 16% in normal bone), its hardness was not much greater than that of normal bone.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

References