

LOOSENING OF A NONCEMENTED POROUS-COATED ANATOMIC FEMORAL COMPONENT IN GAUCHER'S DISEASE A CASE REPORT AND REVIEW OF LITERATURE

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There are few reports of experiences with hip arthroplasty in patients with Gaucher's disease. Review of the literature shows a high rate of loosening after any type of arthroplasty. The clinical, roentgenographic and histopathological findings of a porous-coated hip arthroplasty in Gaucher's disease necessitating revision after 5 years are reported. Because we found typical lipid-laden Gaucher cells at the bone prosthesis interface we support the hypothesis that loosening in Gaucher's disease is due to continued Gaucher cell proliferation and erosion of bone. Loosening seems to be related to the disease and not to a specific type of prosthetic component.

Keywords : Gaucher's disease ; prosthetic loosening ; noncemented arthroplasty.

Mots-clés : Maladie de Gaucher ; descellement de prothèse ; arthroplastie sans ciment.

Gaucher's disease is an inborn error in metabolism characterized by the storage of glucocerebrosides in massively distended reticuloendothelial cells throughout the body, mainly in the liver, the spleen and bone marrow. The deficiency of the glucocerebrosidase enzyme is transmitted as a mendelian autosomal recessive disorder.

In the pediatric types II and III, neurological complications are predominant, whereas in the adult type I, mainly orthopedic complications become apparent (2, 15).

This nonneuronopathic or type I Gaucher's disease is highly variable in both its severity and progression. Several different radiologic manifestations of skeletal lesions are reported : osteolysis, medullary expansion and sclerosis in long bones,

bone collapse, osteitis (4), fractures (10) and secondary deformity can complicate this disease (3).

The most common orthopedic problem for Gaucher's patients is disability due to avascular necrosis of the femoral head (11). Although prosthetic joint replacement is recommended as the treatment of choice in appropriate circumstances, long-term results are variable (12). Moreover few experiences with noncemented hip replacement in patients with Gaucher's disease have been published (1, 8, 9, 11, 13).

We report one case where a noncemented porous-coated anatomic femoral component became loose and subsided four years after implantation. This case is remarkable in its clinical, radiological and histological appearance.

CASE REPORT

An active 47-year-old male engineer presented in January 1986 with increasing and incapacitating pain in the left hip of one year's duration. Mobility of the hip joint was decreased in all planes. His Harris hip score at that time was 87 points. The roentgenographs showed advanced avascular necrosis of the femoral head with collapse of the anterosuperior part. The acetabulum seemed fairly

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intact. Since no joint-preserving surgery was possible for this young patient, he was scheduled for a noncemented arthroplasty.

The general physical examination revealed hepatosplenomegaly, and the routine preoperative screening demonstrated thrombocytopenia with 90,000 platelets/ml. The patient himself did not recall any hemorrhagic diathesis.

A hematological disorder was suspected, and a sternal biopsy with bone-marrow aspiration was performed, showing abundant Gaucher's cells. The demonstration of beta-glucosidase deficiency in peripheral blood leucocytes (5) confirmed the diagnosis of Gaucher's disease.

In February 1986, a hemiarthroplasty (fig. 1) was performed, since during surgery no damage to the acetabular cartilage was noticed. Therefore,



Fig. 1. — Hemiarthroplasty with a noncemented P.C.A. stem and bipolar head.

only the femoral head and neck were resected and replaced by a prosthesis using a PCA (Howmedica, Rutherford, New Jersey) noncemented stem in combination with a bipolar head (Depuy, Warsaw, Indiana). Blood loss was within standard limits, and the postoperative course was uneventful.

Yearly radiographical and clinical examinations were excellent with Harris hip ratings of 96 at one, two and three years postoperatively. In January 1990 however the patient started complaining of mild pain in the mid thigh. Roentgenograms revealed focal endosteal scalloping around the smooth part of the prosthetic stem.

Four months later he presented with a Harris hip score of 55 points, and the roentgenographs showed an unstable prosthesis. The femoral component had subsided several centimeters in the osteopenic femoral shaft. Loose beads were scattered at the bone-prosthesis interface and endosteal cavitation had appeared since the previous x ray (fig. 2). The patient was scheduled for revision surgery. Prior to surgery epicutaneous patch testing showed no allergy to the metal components of the alloy.

During revision, the grossly loose prosthesis could be very easily extracted by hand. A well-organized fibrous membrane without evidence of metallosis surrounded the stem and was well anchored to the porous-coating. An intraoperative hip aspirate was sterile. The femoral shaft was thoroughly curetted until a bleeding bone surface was reached. Morcellized cancellous allografts were placed into the proximal femur and packed together using the standard rasps. Thereafter additional impaction of the grafts was obtained by hammering in a noncemented hydroxyapatite-coated Profile stem (DePuy, Warsaw, Indiana).

Since there was no visible evidence of wear of the acetabular cartilage, the acetabulum was left untouched and a bipolar head was placed on the femoral stem.

Postoperatively the patient was placed in traction-suspension for 6 weeks. No weightbearing was permitted for 6 more weeks thereafter.

Histological examination of the fibrous membrane showed densely organized collagenous tissue packed with Gaucher's cells (fig. 3). These cells are typically characterized by the eccentrically



Fig. 2. — Unstable femoral component subsided in the osteolytic femur.

located nucleus, and the expanded cytoplasm appearing vacuolated. Under polarized light, only a few scattered and irregular refractive bodies were visible. There was no evidence of malignant disease or infection. No microorganisms were cultured from this tissue.

Two years after revision hemiarthroplasty the patient is doing well, with a Harris hip score of 82 points and a roentgenographically stable femoral component.

DISCUSSION

The treatment of the affected hip in Gaucher's disease has been discussed by several authors

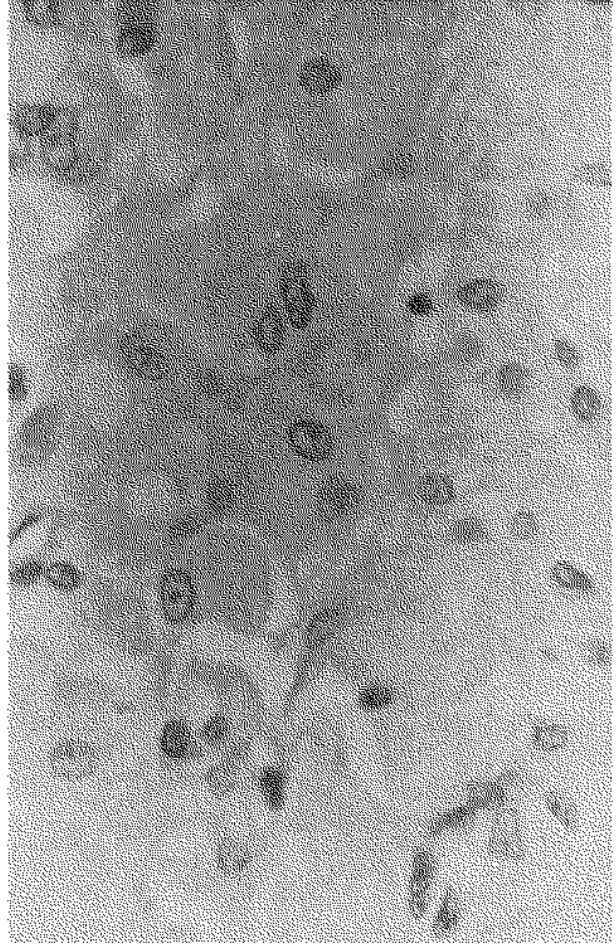


Fig. 3. — Biopsy of the bone-prosthesis interface shows abundant Gaucher's cells. (Hematoxylineosin $\times 250$).

(1, 3, 8, 11, 13). All authors agree that joint arthroplasty restores satisfactory joint mobility to Gaucher patients who were incapacitated prior to the operation. Aseptic loosening of the prosthetic component however remains a problem (12). Implant failure has been observed after uncemented Austin-Moore hemiarthroplasty and double cup arthroplasty as well as cemented total hip arthroplasty. Some authors (1, 13) reported good results with resurfacing arthroplasties, a procedure which might now be considered as obsolete (7). Amstutz and Carey (1) treated four of their patients with a noncemented Austin-Moore hemiarthroplasty; the short-term results, after a follow-up of less than 18 months were good. Herndon and Aufranc

(9) reported a good result in one case 4 years after a similar procedure.

Longer follow-up studies of other hemiarthroplasty series in Gaucher's disease are not available.

Osteolytic changes in the upper femoral shaft after implantation of a porous-coated femoral component for indications other than Gaucher's disease are well known (6, 14). Hypersensitivity to metal, or foreign body response to polyethylene or metallic wear debris were quoted as possible causes of the lysis (16). In our case the negative epicutaneous patch testing as well as the absence of many lymphocytes or birefringent particles at the site of bone resorption do not support this hypothesis.

Lipid-laden cells, however, were abundantly present at the bone-prosthesis interface. This supports the hypothesis according to which the greater tendency for implant failure in Gaucher's disease is due to continued Gaucher's cell proliferation. The biological signals responsible for the accumulation of Gaucher's cells in this region of the femur are poorly understood. The bone of Gaucher patients might just simply react to the presence of a foreign body causing changed stress patterns in the proximal femur.

Although the pathogenesis remains obscure, this case as well as the review of the literature clearly shows that prosthetic femoral components in Gaucher patients are more prone to aseptic loosening. Our findings indicate that, at least on the femoral side, this phenomenon seems to be more related to the disease itself, than to a specific geometric design, a specific alloy, the surface finish of the prosthetic component or the use of cement.

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SAMENVATTING

P. A. J. VAN WELLEN, P. HAENTJENS, N. FREYCOURT, P. OPDECAM. Loslating van een niet-gecementeerde PCA prothese bij ziekte van Gaucher.

De resultaten van de heuparthroplastiek bij patiënten met de ziekte van Gaucher zijn onvoldoende gekend. De weinige publicaties hiéover vermelden een snelle loslating van om het even welk type arthroplastiek. De klinische, radiografische en histologische bevindingen worden weergegeven bij een patiënt met de ziekte van Gaucher en met loslating van een niet-gecemen-

teerde PCA-prothese. Deze prothese werd gereviseerd na 5 jaar follow-up wegens inzakking van de femorale component.

Wij vonden typische cellen van Gaucher ter hoogte van het overgangsgebied tussen prothese en bot. Dit ondersteunde onze hypothese dat de loslating het gevolg kan zijn van de voortdurende proliferatie van cellen van Gaucher en boterosie. Het type van heupprothese lijkt de prognose van de arthroplastiek bij patiënten met de ziekte van Gaucher niet te beïnvloeden.

RÉSUMÉ

P. A. J. VAN WELLEN, P. HAENTJENS, N. FRE-COURT, P. OPDECAM. Descellement aseptique d'une prothèse fémorale non-cimentée de type PCA chez un patient atteint de maladie de Gaucher.

La littérature met en évidence un taux élevé de descellements aseptiques précoces chez les patients atteints

de maladie de Gaucher, quel que soit le type d'implant utilisé.

Nous rapportons les données cliniques, radiologiques et histologiques d'un patient atteint de maladie de Gaucher ayant bénéficié d'une révision de la tige fémorale d'une prothèse non-cimentée de type PCA.

L'étude histologique de l'interface implant fémoral-os a montré la présence de multiples cellules typiques de la maladie de Gaucher. Ces données histologiques confirment l'hypothèse selon laquelle une prolifération de cellules de Gaucher pourrait être la cause réelle de l'ostéolyse et du descellement aseptique chez ces patients, quel que soit le type de prothèse utilisée.