Case Report

Total hip arthroplasty in Hurler syndrome – 8 years follow up – A case report with review of literature

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ABSTRACT

Life expectancy in Hurler syndrome is significantly improved by enzyme therapy with bone marrow transplantation. However, the deterioration of skeletal abnormalities persists. Hip dysplasia is a common presentation which may progress to significant hip arthritis requiring total hip arthroplasty at later stage. We report a long-term outcome of cementless total hip arthroplasty in a patient with Hurler syndrome who was successfully treated with bone marrow transplant.

1. Case report

A 17-year-old female, known case of Hurler syndrome and successfully treated with Bone Marrow Transplantation had presented to us 8 years back with complaint of pain in both the hips for one year duration. The pain was more in left hip as compared to the right. On examination, movements in the left hip were painfully restricted with flexion, abduction and external rotation being 0–50°; 0–10° and 0–5° respectively. In the right hip, the movements were restricted but relatively less painful with flexion 0–90°, abduction 0–15° and external rotation 0–15°. However; extension, adduction and internal rotation were restricted in both the hips. On radiographic evaluation, there was reduction in joint space in both the hips (Fig. 1). Considering the clinical presentation, and the fact that she was more symptomatic on the left side, she was counselled for total hip arthroplasty on the left side only. The surgery was performed via standard posterior Moore's approach. The uncemented components (Pinnacle® Acetabular shell size 44 mm, Pinnacle™ metal insert 28 mm ID × 44 mm OD and Proxima™ Hip Duofix stem size 1; Depuy International, Leeds, UK) were used. Considering her young age, the short stem prosthesis was used. She had a small split 1 cm long in proximal femur while inserting the smallest size stem which was fixed with cerclage wiring. Sutures were removed at 2 weeks. The patient was advised gradual mobilization from toe-touch weight bearing initially to partial weight bearing at 6 weeks and then full weight bearing by 3 months.

She was followed up every month for first 3 months and then every three months for the next six months. Thenafter she was scheduled for follow up at one year followed by a yearly follow up. On each visit, she was assessed clinically. Surprisingly, her intensity of pain decreased on

the right side after she underwent surgery on the left side. Apart from the immediate postoperative period, radiographs were taken at 3 months and at one year to look for implant position, both of which revealed stable implants in situ. Thenafter the radiographs were taken yearly on each visit which too revealed stable implants. She used to complain of occasional aggravation of pain in right hip at times during the follow up but could get relief with low dose analgesics, quite often with paracetamol 500 mg only as and when required. At the latest follow-up of 8 years, she is doing well with her left hip and can do her activities of daily living without any difficulty. She has a limb length discrepancy of 2 cm which she is managing with a shoe raise on her right side. She is still occasionally having pain in her right hip but is not abusing any analgesics. She has been informed regarding the possible need of total hip arthroplasty in right side in future. The latest X-ray shows the stable implants in left hip without any signs of loosening or failure (Fig. 2). As she had a metal on metal articulation, she is strictly under regular follow up pertaining to her left hip. The latest blood ions levels were within the safe range (Cobalt = 4.71 µg/L, Chromium = 25.06 µg/L). Furthermore, there was no collections or any abnormalities seen in local ultrasonography or in metal artefact reduction sequence (MARS) magnetic resonance imaging done as a part of thesis by a post graduate trainee at our institute. Currently she is also under regular follow up in Gastroenterology department for her liver ailments.

2. Discussion

Hurler syndrome (Mucopolysaccharidosis type I) is an autosomal recessive disorder leading to the accumulation of glycosaminoglycans

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Fig. 1. X-ray of both hips with pelvis showing reduction of joint space and irregularity of joint margins in both the hips. Note the deformed femoral head.

Fig. 2. X-ray of both hips with pelvis showing stable total hip prosthesis in left side. Note the cerclage wire which was applied to fix the crack encountered during implantation of femoral stem.

in all tissues. This results from a deficiency of the enzyme α -L-iduridase resulting in inability to break down the glycosaminoglycans which consequently accumulate in the tissues.^{1,2} The array of musculoskeletal abnormalities in Hurler syndrome includes hip dysplasia, knee deformity, kyphosis of the spine, atlantoaxial instability and joint stiffness. Hip dysplasia occurs due to failure of ossification of the lateral acetabular roof, medial proximal epiphysis of the femur both leading to a dysplastic hip which is often accompanied by deformation or dislocation and avascular necrosis of the femoral head.³

Enzyme therapy with bone marrow transplantation (BMT) has been employed as a successful treatment for Hurler syndrome. The life expectancy is increased with enzyme therapy as the systemic abnormalities are improved.⁴ However, even after successful BMT, the overall progression of skeletal disease is significant. Furthermore, studies have shown that the outcomes are poor in lower extremities.^{5,6} Most authors advocate early surgical intervention i.e. containment osteotomies in patients who do not have arthritic symptoms.⁷ However, in long term survivors, the hip dysplasia may progress to hip arthritis that warrants the necessity of total hip arthroplasty. Satisfactory results have been reported following total hip arthroplasty in patients with other types of Mucopolysaccharidosis but there is paucity of literature regarding total hip arthroplasty in patients with type I (Hurler syndrome). The available reports have published short term follow up only. This is the first

report to describe the long-term outcome of total hip arthroplasty in a patient with Hurler Syndrome.

O'hEireamhoin et al⁸ reported a 21-year-old female with Hurler syndrome post bone marrow transplant at the age of 20 months, who underwent total hip replacement in her left hip using cementless prosthesis. At 2 years follow up, she had recovered well and returned to her previous level of assisted community ambulation. Ledford et al⁹ published a series of 5 very young bone marrow transplant patients, one of them with Hurler syndrome, who underwent bilateral sequential total hip arthroplasty. The patient had a history of multiple pelvic osteotomies; subsequently the total hip arthroplasty was challenging. There was a periprosthetic fracture around the femur intraoperatively which was fixed with two cerclage wires and strut allografts. The patient had a re-fracture due to fall, 8 months following the hip arthroplasty which was again fixed with plate and strut allografts. At the follow up of 50 months, the patient had a good self-reported outcome. In the current case also, the authors encountered an intraoperative split spanning 1 cm in the proximal femur while implanting the smallest size available prosthesis suggesting the poor quality of bone and narrow medullary canal in such patients which cautions to be careful while performing prosthetic replacement. Kennedy et al¹⁰ retrospectively reviewed the long term functional and radiological outcome of pediatric hip surgeries in patients with Hurler syndrome and reported that 10 out of 26 hips had radiologic degenerative changes despite stable well covered hips. They concluded that the active intervention at early age does not prevent the development of radiologic deterioration of hips and hence recommended that the pediatric hip surgery should be designed with the possibility of early total hip arthroplasty in mind in patients with Hurler Syndrome.

As the life expectancy of the patients with Hurler syndrome is considerably increased after BMT, total hip arthroplasty may be an important treatment option for those with symptomatic osteoarthritis of the hips in whom the corrective surgery is not likely to be effective in relieving the symptoms. Though we don't recommend a metal on metal prosthesis at present due to growing problems and recall, cementless prosthesis may be a reliable option; however, precautions should be taken to prevent any periprosthetic fractures. Long term satisfactory clinical and radiological results can be expected.

Conflict of interests

None.

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