On-line Case Report

Total hip arthroplasty in Klippel–Trenaunay syndrome

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ABSTRACT

Klippel–Trenaunay syndrome patients often suffer degenerative joint disease at an early age. Performing arthroplasty in these patients is particularly difficult for a number of reasons. In this case report, we describe the second reported case of total hip replacement in Klippel–Trenaunay syndrome, using novel techniques to surmount the problems faced in this challenging condition.

Keywords: Klippel Trenaunay syndrome – Arthroplasty – Hip – Sclerotherapy – CAD-CAM

Klippel–Trenaunay syndrome is a congenital disorder first described in 1900 by the French physicians Klippel and Trenaunay.¹ They described a syndrome characterised by cutaneous vascular naevus, soft tissue and bony hypertrophy, and venous malformations. Typically, only one limb is involved, with the lower limb representing 70% of cases.

Klippel–Trenaunay syndrome is frequently a painful condition, with degenerative joint disease being one of the many potential causes of pain. There are several underlying causes for degenerative joint disease including leg-length discrepancy, vascular malformations within the joint leading to recurrent haemarthrosis, or intra-osseous vascular malformations leading to abnormal bony architecture.²

Standard practice states that bony surgery should be avoided in Klippel–Trenaunay syndrome at all costs due to the risks of uncontrollable haemorrhage. A review of the literature regarding arthroplasty in Klippel–Trenaunay syndrome revealed only one reported case of total hip arthroplasty in a patient without bony involvement, and one of total knee arthroplasty where a tourniquet could be used.^{3,4} Both reports highlight the difficulties of performing arthroplasty in this patient group. We report a further case

of total hip arthroplasty in a 27-year-old woman with significant bony involvement, including the application of a variety of novel strategies to minimise complications in this technically demanding procedure.

Case report

A 27-year-old woman presented with worsening left hip pain over the past 5 years. The pain was constant and was significantly limiting her activities. Her past medical history was of Klippel–Trenaunay syndrome with left leg hypertrophy. She was thought to have had a premature growth arrest of the femoral epiphysis secondary to a spontaneous haemarthrosis, and had undergone a subtrochanteric femoral shortening osteotomy as a child. Examination revealed a hypertrophic left leg, with multiple superficial varicosities. Leg lengths were equal with a short left femur, and long left tibia. Range of movement of the hip was significantly reduced and painful.

Plain radiographs revealed marked abnormality of the left hip with a short femoral neck, a flattened femoral head, and multiple bony defects within the proximal femur, which corresponded to substantial vascular

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Figure 1 Arteriography showing intra-osseous vascular channels, and postoperative radiograph showing CAD-CAM prosthesis in situ.

channels. The acetabulum was shallow, and the joint showed degenerative changes.

Arteriography was used to assess the aberrant vasculature. This confirmed multiple intra-osseous, high-flow, arteriovenous malformations corresponding to the defects on plain radiographs with diameters up to 2 cm (Fig. 1).

Because of the severity of the patient's symptoms, the decision was made to proceed to total hip arthroplasty.

Due to the abnormal morphology of the proximal femur, a custom-made femoral prosthesis was used. A fully hydroxyapatite-coated femoral stem was produced using CAD-CAM technology based on the CT scans and calibrated plain radiographs (Stanmore Implants Worldwide). This type of femoral component has been shown to have good outcomes in young adults in inflammatory polyarthropathy, which has traditionally been regarded as a challenging area.⁵

Relatively normal acetabular morphology permitted the use of a standard uncemented acetabular component (JRI Furlong HAC CSF plus acetabular cup). Due to the age of the patient, a 32-mm diameter ceramic-on-ceramic, large diameter articulation was selected for longevity.

In order to minimise intra-operative blood loss from the intra-osseous vascular malformations, pre-operative embolisation was employed. Under general anaesthesia, angiography of the internal iliac, external iliac, circumflex femoral and profunda femoris arteries was performed. Extensive abnormal vascularity with rapid arteriovenous shunting into grossly dilated veins was demonstrated in the femoral head and neck. Draining veins were punctured percutaneously and embolised using Histoacryl immediately adjacent to the arteriovenous communications, resulting in complete obliteration of

arteriovenous shunting. To reduce allogenic blood transfusion requirements, a cell saver (Hemonetics; Braintree, MA, USA) was used intra-operatively and salvaged blood re-transfused.

The arthroplasty was carried out as planned via the posterior approach. Significant bleeding was encountered from grossly abnormal vessels within the soft tissues, which was controllable with ligation and cautery. However, there was no significant bony bleeding and the bone quality was excellent.

Salvaged autogenous blood of 2.1 l was re-transfused during the procedure. Haemoglobin was 11.3 g/dl pre-operatively and dropped to 9.2 g/dl postoperatively. The patient experienced no adverse events, was mobilizing on the second postoperative day and discharged home on the fifth postoperative day. At 6-month follow-up, the patient was walking with a normal gait, no discomfort and was back to all desired activities.

Discussion

Patients with Klippel–Trenaunay syndrome undergoing arthroplasty pose significant challenges due to both the abnormal bony morphology and the abnormal vasculature that places them at great risk of uncontrollable haemorrhage.

Intra-osseous malformations are rare, but it is critical to recognise their presence. In this case, had these not been recognised and embolised pre-operatively, there would have been a high risk of exsanguination. Despite this, we still encountered significant bleeding from the soft tissues which required extensive cautery and ligation. The use of a cell saver allowed a reduction in requirements for allogenic transfusion; however, a significant quantity of allogenic blood was still required.

Mallick and Weeber³ noted intense venous bleeding despite the use of adrenaline at the incision site, and the normal appearance of vasculature on pre-operative MRI. Their case differed from ours in that their patient did not have intra-osseous malformations. Catre *et al.*,⁴ operating on the knee, were able to use a tourniquet and, hence, avoided significant haemorrhage; however, they noted many haemangiomas and varicosities.

The abnormal shape of the proximal femur as a result of the condition and the previous surgery meant that durable fixation of a conventional femoral component would be difficult. This was addressed by using a custom-made implant.

Conclusions

With this case, we have used modern techniques in order to surmount the problems posed by Klippel–Trenaunay

syndrome. We would advocate the consideration of these techniques when arthroplasty is envisaged in patients with Klippel–Trenaunay syndrome or in other challenging situations.

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