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Total Hip Replacement in Klippel-Trenaunay Syndrome

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Abstract

Klippel-Trenaunay syndrome (KTS) is very rare and severe degenerative joint disease at an early age. Orthopedic intervention in these patients brings numerous difficulties for various reasons. Very few cases of total hip replacement in KTS have been documented in the literature. We present this case report, where a twenty-five-year male was presented with pain in left hip for the last eighteen months. Our contemporary method to conquer the situations dealt with this keen condition. Acetabular morphology was normal and also the bone quality was good enough, which permitted the use of a standard acetabular as well as femoral component. Non cemented acetabular and femoral component with large diameter articulation was selected. Pre-operative embolization was employed before forty-eight hours of surgery to minimize per operative blood loss from the intra-osseous vascular malformation; besides, a cell saver was also used pre-operatively, which helped to reduce allogeneic blood transfusion requirements. Significant bleeding was encountered per operatively from grossly abnormal vessels within the soft tissues, which was controllable with ligation and cautery. The patient was discharged on the fifth day of surgery; first follow-up was done at two weeks of surgery after complete healing of the wound. At sixteenth week follow-up, the patient was walking with a normal gait and was back to all desired activities.

Introduction

Klippel-Trenaunay syndrome (KTS) was first coined in 1900 by the French physicians Klippel and Trenaunay [1]. It is a congenital disorder consisting of cutaneous vascular nevus, soft tissue and bony hypertrophy and venous malformations [2]. The etiology of KTS still remains obscure [3]; usually one limb is involved, with the lower limb representing 70% and upper limb almost 11% of cases [4]. Pain around the joint in this syndrome is frequently crippling. It is associated with a degenerative joint disease which is one of the potential causes of pain. In this syndrome, leg-length discrepancy, vascular malformations within the joint causing recurrent haemarthrosis or intra-osseous vascular malformations lead to abnormal bony changes [5].

The standard practice advocates, skeleton surgery should often be avoided in KTS, due to the risk of uncontrollable hemorrhage. The previous studies regarding arthroplasty in KTS concluded advantages of a tourniquet in total knee replacement, which decreased bleeding during the procedure [6, 7]. We report a case of total hip arthroplasty in twenty-five years male with bony involvement and significant pain.

Case report

A twenty-five-year male was presented with pain in left hip for the last eighteen months. The pain had been continuous and used to aggravate while walking, but recently, it started limiting daily activities. Examination revealed varicosities in left groin down to the penile region, including the anterior abdomen (Fig. 1). Leg length discrepancy was not found. The range of movements of the hip was significantly reduced and painful. Plain radiographs revealed avascular necrosis of femoral head (Fig. 2).

The patient was planned for surgery and further work-up was done as per routine procedure for total hip replacement. Doppler study was considered in this case. Varicosities were pronounced all over the left lower limb but no deep vein thrombosis was noted. Further CT venogram was done seeing the

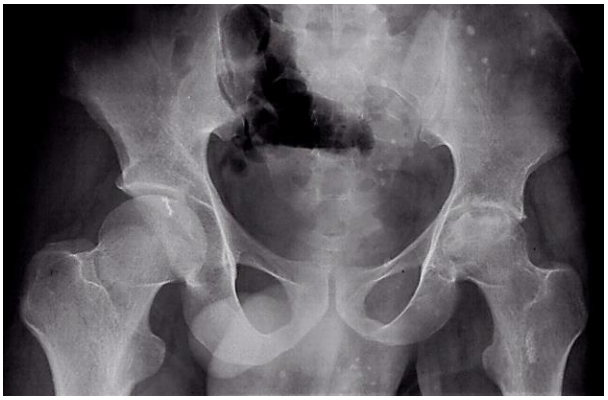

Fig. 1 Radiograph showing avascular necrosis of the femoral head.

results of color-Doppler, which showed the same results. Surgery was planned along with vascular surgeons, the patient was admitted three days prior to surgery and posterior approach was considered. Considering the severity of the patient's symptoms, the decision was made to proceed for total hip arthroplasty. Under combined epidural and spinal anesthesia operation was planned, a posterior approach was used for total hip replacement. The proximal part of the approach was free of vascular anomalies, but just distal to the tip of the trochanter, engorged vessels were present. Significant bleeding was encountered from grossly abnormal vessels; approximated bleeding during surgery was estimated to be two liters. However, there was no significant bony bleeding and the bone quality was good. Controlling blood loss was a challenge as it was difficult to ligate or cauterize the varicose vessels. The bleeding was stopped and the preparation for the acetabular and femoral component was done (Fig. 3). This further aggravated the bleeding from the bone and from each ligated distal varicosities. The femoral component was implanted then bleeding was controlled. The total amount of per-operative blood loss was approximated to be two liters. Closure over the drain was done. Around 800 ml of blood was transfused per operatively, the same amount of blood was transfused in the post-operative period and the patient was discharged on fifth days of surgery, first follow-up was done at two weeks of surgery after complete healing of the wound. At sixteenth week follow-up, the patient was walking with a normal gait and was back to all desired

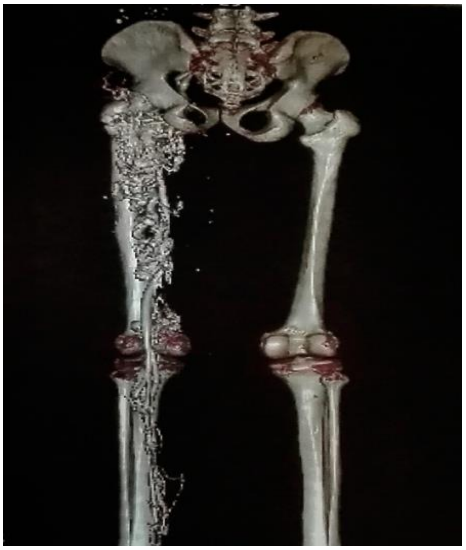

Fig. 2 CT venogram, abnormal varicosities in the left lower limb.

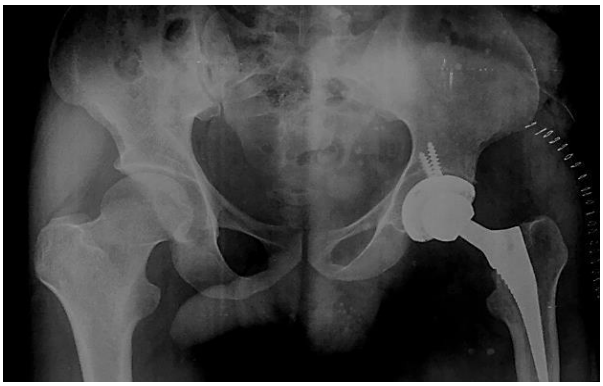

Fig. 3 Postoperative radiograph showing prosthesis *in situ*.

activities.

Discussion

Klippel-Trenaunay syndrome is a rare syndrome and only a few reports of total joint have been mentioned in the literature following this syndrome. It is of utmost importance to plan preoperatively in such condition, due to challenges of massive potential bleeding faced during surgery and dislodgement of thrombus after surgery, which can be associated with this syndrome since past [8]. The approach should be determined by prior evaluation through CT angiogram, through a minimal area of varicosities to reduce blood loss [9]. Thus, extensive work-up before surgery, selection of surgical approach and planning for the blood loss during surgery has to be planned with the team effort of an orthopedic surgeon, vascular surgeon, anesthesiologist and physician [10]. There are only a few case reports of primary replacement in the literature and difficulty with revision surgery is yet to be seen.

Conclusions

In this case, we had a proper preoperative evaluation and planning to conquer the problems posed by KTS. We would advocate the consideration for proper preoperative evaluation, team approach, including vascular surgeon, radiologist and anesthesiologist along with the adequate availability of blood when arthroplasty is contemplated in patients with KTS or in other challenging situations like preparation and selection of an acetabular and femoral component.

Conflict of interest

The authors declare that they have no conflict of interest.

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