Case Report

Total knee arthroplasty in vascular malformation

Harish Bhende, Nanadkishore Laud, Sandeep Deore, V Shashidhar

Abstract

In Klippel–Trenaunay syndrome, vascular malformations are not only in skin and superficial soft tissues but also in deep tissues like muscles bones and joints. It is well documemted that these recurrent intraarticular bleeds can cause early arthritis and joint pain. Performing arthroplasty in such patients is difficult and fraught with complications. We describe such a case where navigated total knee arthroplasty was performed with success to avoid the problems of intra medullary alignment used in the presence of intra medullary vascular malformations. We also suggest certain measures when knee arthroplasty is considered in such patients.

Key words: Knee arthroplasty, navigation surgery, vascular malformation
MeSH terms: Arthroplasty, replacement, knee, surgery, computer assisted, vascular neoplasms

Introduction

Vascular malformation of a unilateral lower limb with severe arthritis of the same knee joint is a rarity. Total knee arthroplasty (TKA) in such case is fraught with complications. Baskerville has reported 16% rate of deep vein thrombosis (DVT) and 14% risk of pulmonary embolism in patients with Klippel–Trenaunay syndrome.[1-3] Same view is endorsed by Muluk *et al*.[4] Silva and Luck have reported high rate of late infections with 10-year survival of infection free implants being 77% in hemophilic knees undergoing TKA.[5] We report a case of early onset degenerative knee arthritis with vascular malformations in which navigated TKA was performed successfully. Only two cases are reported in the literature, which have some resemblance to our case.[6,7] Our case is unique to have intramedullary lesions in the femur to complicate the management.

Case Report

A 30-year-old woman working as a clerical staff, requiring daily commuting by crowded trains, was suffering from left knee pain for last 6 years. Her difficulty in walking had worsened since last 6 months and did not respond to nonsurgical management. She was bedridden for 2 months before she presented to us. She also had a history of intermittent rectal bleeding.

On examination, there were cutaneous and soft tissue hemangiomas alongwith varicose veins throughout the affected left lower extremity from hip to toes, mainly on the gluteal region, posterior thigh, popliteal fossa, calf and foot [Figure 1a-d]. There were no visible salmon patches or portwine stains. Left lower extremity was not significantly enlarged compared to right lower extremity. There were no lesions over the anterior aspect of left knee. There was a 10° valgus alignment of the knee [Figure 2]. She had a flexion deformity of 20° with further flexion up to 40°. The movements were very painful in the available range of motion (ROM). The knee society pain score was 29 and her functional score was 0. There was no true leg length discrepancy. Dorsalis pedis and posterior tibial pulsations were palpable. The distal neurological status was normal. The right knee had full range of movements with no pain or deformity. There were no vascular lesions in right lower extremity. Bilateral hip examination was normal.

Radiographs [Figure 2] showed severe degenerative tricompartmental arthritis in the left knee joint with bone loss from the lateral femoral condyle and the lateral tibial plateau. There was subchondral sclerosis and osteophyte formation. Phleboliths in the vascular malformations were noticed on the X-rays.

Magnetic resonance imaging and magnetic resonance venogram [Figure 3] of the left lower limb with pelvis showed vascular malformation; mostly multifocal hemangiomas

Center for Joint Replacement Surgery, Laud Clinic, Saraswati Nilayam, Dadar East, Mumbai, Maharashtra, India

Address for correspondence: Dr. Harish Bhende,
Center for Joint Replacement Surgery, Laud Clinic, 180, Saraswati Nilayam, Hindu Colony, Dadar East, Mumbai - 400 014, Maharashtra, India.
E-mail: harishbhende@gmail.com

Access this article online	
Quick Response Code:	Website: www.ijoonline.com
	DOI: 10.4103/0019-5413.164049

Figure 1: Clinical photographs of patient showing (a-d) The extent of superficial vascular lesions from left hip to toes with flexion contracture of the left knee

Figure 2: (a and b) X-rays of left knee anteroposterior and Lateral views showing severe degeneration and the phleboliths (arrow)

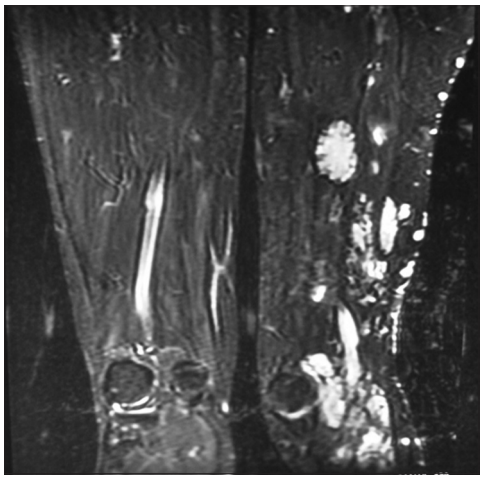

Figure 3: Magnetic resonance venogram showing vascular malformation in intra medullary, subcutaneous and deep soft tissues with phleboliths

involving superficial and deep soft tissues of the entire left lower limb, pelvic region, rectum and left femur. There were no communications between the superficial and the deep lesions. Dual phase angiogram did not show any feeder artery or draining vein. As there were no feeder vessels, preoperative embolization was not done. Her coagulation profile was normal (bleeding time -5 s, clotting time -3 min, prothrombin time-13 s, international normalization ratio-1.1, activated partial thromboplastin time-28 s, Factor VIII-110% and Factor IX-100%). She did not have any systemic involvement. In the absence of limb hypertrophy, we could not label it as classical Klippel–Trenaunay syndrome, but it can be called as a variant of Klippel–Trenaunay syndrome.

Surgery was done under spinal anesthesia[8] with tourniquet and exsanguination of limb using Eshmarch bandage. Anterior midline incision with medial parapatellar approach was used. Computer-assisted navigated surgery was done due to intra medullary involvement of the lesions [Figure 3]. Ci navigation system (DePuy, Brainlab, Germany) was used. Two thin threaded pins with bicortical holds were used to fix the clamps carrying the tracker ball arrays. The pin hold was good and remained well till the end of the surgery as the bone quality was good on both femoral as well as tibial sides. The synovium was grey-black with an orange tinge of the remaining articular cartilage. Femoral condyles were squared. Femoral bone cuts also revealed vascular malformations in the bone which were curetted out [Figure 4].

Cemented TKA using posterior cruciate substituting femoral prosthesis (PFC Sigma, Depuy, Johnson and Johnson) with all-poly tibial component [Figure 5] was done. Bone wax was used to control bony bleeding from metaphyseal lesions at the femoral cuts. Complete hemostasis was achieved after release of tourniquet. Closure was done under re-inflated tourniquet as there was generalized ooze but no active bleeding. Total tourniquet time was 80 min. Intravenous (I.V) tranexamic acid was used (900 mg I.V. infusion, first dose 30 min before tourniquet inflation, second dose 3 h later, and additional third dose 6 h after second dose).

Postoperatively, her hemoglobin dropped from 12.5 g% to 8.6 g% and she was transfused three units of packed cell volume. Intra-operative blood loss was 300 ml. Postoperative drain was kept for 48 h. The total collection in drain was 650 ml. Prophylactically oral rivoroxaban 10 mg (Xeralto - Bayer Zydus Pharma) was given 2 weeks postsurgery, because such patients have increased risk of thromboembolism.[3,4] Subsequently, she was kept on oral aspirin 75 mg daily for a period of 4 weeks. Patient was kept on intermittent calf pumps for 3 days postsurgery. She was given DVT above knee stocking and encouraged to do active ankle-foot exercises. Aggressive mobilization was

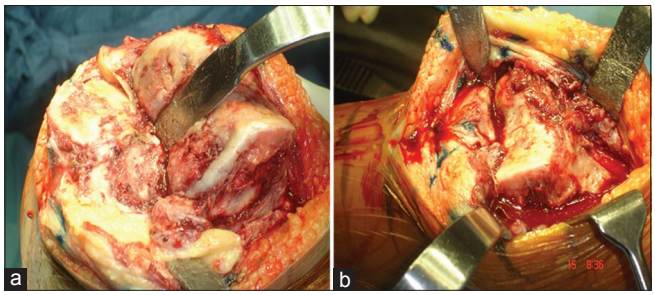

Figure 4: (a and b) Intra operative photographs showing the synovium, femoral condyles and cut distal femur

used under supervision of a physiotherapist. Continuous passive motion and full weight bearing walking with a walker were started from next day of surgery. She was walking with walker support and had 0–90° ROM by 7th day, when she was discharged. Her wound healed well, and sutures were removed on the 15th day postsurgery.

At 1-year followup, her knee society pain score improved to 97 and functional score improved to 100. She was walking normally without pain and had joined her job. She was also able to commute by crowded public trains without any support.

Histopathological examination of the removed bone and synovial tissue showed vascular proliferation with lymphoplasmacytic exudates. Conglomerate of vessels with thick walls and red blood cells in slit like lumina with hemosiderophages were seen. These findings were consistent with the histological examination of hemangiomas of Klippel–Trenaunay Syndrome.[7]

Discussion

Klippel–Trenaunay syndrome is a type of arterio-venous malformation with three features: Cutaneous hemangiomas, limb hypertrophy and varicose veins. They can also have per rectal bleeding. Vascular malformations are polysystemic in nature.[9]

Our patient had all of the above except limb hypertrophy, so we can call it as a variant of Klippel–Trenaunay syndrome. Unilateral knee arthritis on the side of vascular malformations probably happened due to recurrent chronic bleeds in her knee from synovial hemangiomas. These repeated minor bleeds gave the knee gross appearance similar to a hemophilic knee. The radiological squaring of femoral condyles and histologic presence of hemosiderophages made it to look like hemophilic arthritis (although her coagulation profile was normal).

After the TKA, there is still future risk of recurrent bleeds in her left knee due to persistent hemangiomas in the

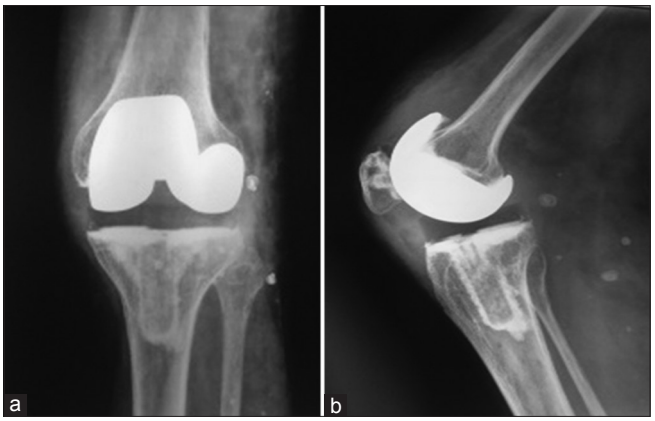

Figure 5: Postoperative radiographs anteroposterior (a) and lateral (b) views showing good alignment. All-poly tibial component was used

synovium increasing the risk of infection (as it is observed in hemophilic knees undergoing TKA).[10] Challenges during surgery include identifying the extent of vascular anomaly and avoiding vascular injury resulting into uncontrolled bleeding. The presence of intra-medullary large lesion in this case precluded the use of intramedullary instrumentation. We had to use navigation technique for arthroplasty.

The only two cases of Kippel–Trenauney syndrome reported in literature, where knee arthroplasty had been performed, did not have intramedullary and intraosseous vascular malformation. The presence of intra-osseous vascular malformation makes the treatment by arthroplasty more difficult as we need to plan to avoid them during our bone cuts.

Our patient was young and her bone quality was good. Recent reports show that there is no significant difference between an all-poly and metal backed tibial component in terms of survival and function.[11,12] The risk of back wear and osteolysis is reduced in all-poly tibia and the revision is easier.[13] Keeping all these in mind, an all-poly tibial component was used in this patient.

We would like to suggest the following while performing knee arthroplasty in the patient with vascular malformation. First, proper vascular studies are essential to assess the intraosseous and deep extent of vascular malformation. Second, vascular surgeon must be consulted if preoperative embolization is required. Third, navigated TKA surgery may be helpful to avoid opening intramedullary space and intraosseous lesions. Fourth, tranexamic acid should be used peri operatively to reduce intra operative blood loss, along with adequate anticoagulation prophylaxis to prevent DVT. Special attention to asepsis must be paid as these patients are probably more prone to infection. The patient should be explained regarding guarded long term

prognosis as the medical condition is un predictable in its progression.

To conclude, TKA in the presence of vascular malformations provides good functional score and better quality life in young patients when done with certain precautionary measures.

References

1. CA Willis-Owen and JP Cobb Total Hip Arthroplasty in Klippel–Trenaunay Syndrome, Ann R Coll Surg Engl. Nov 2008; 90 (8): W6–W8.
2. Lee A1, Driscoll D. Evaluation and management of pain in patients with Klippel-Trenaunay syndrome: A review.Pediatrics 2005 Mar;115 (3):744-9
3. Baskerville PA. Thromboembolic disease and congenital venous abnormalities. Phlebologie 1987;40:531-6.
4. Muluk SC, Ginns LC, Semigran MJ, Kaufman JA, Gertler JP. Klippel-Trénaunay syndrome with multiple pulmonary emboli – An unusual cause of progressive pulmonary dysfunction. J Vasc Surg 1995;21:686-90.
5. Silva M, Luck JV Jr. Long term results of primary total knee replacement in patients with hemophilia. J Bone Joint Surg Am 2005;87:85-91.
6. Leal J, Davies AP, Selmi TA, Neyret P. Knee Arthroplasty in Klippel-Trenaunay syndrome: A case presentation with 5 years followup. J Arthroplasty 2008;23:623-6.
7. Catre MG, Kolin A, Waddell JP. Total knee arthroplasty in Klippel-Trenaunay syndrome. Can J Surg 2005;48:494-5.
8. Holak EJ, Pagel PS. Successful use of spinal anesthesia in a patient with severe Klippel-Trénaunay syndrome associated with upper airway abnormalities and chronic Kasabach-Merritt coagulopathy. J Anesth 2010;24:134-8.
9. Garzon MC, Huang JT, Enjolras O, Frieden IJ. Vascular malformations. Part II: Associated syndromes. J Am Acad Dermatol 2007;56:541-64.
10. Miller MD. Review of Orthopaedics. 2nd ed. Toronto: WB Saunders; 1996.
11. Nouta KA, Verra WC, Pijls BG, Schoones JW, Nelissen RG. All-polyethylene tibial components are equal to metal-backed components: Systematic review and meta-regression. Clin Orthop Relat Res 2012;470:3549-59.
12. Voigt J, Mosier M. Cemented all-polyethylene and metal-backed polyethylene tibial components used for primary total knee arthroplasty: A systematic review of the literature and meta-analysis of randomized controlled trials involving 1798 primary total knee implants. J Bone Joint Surg Am 2011;93:1790-8.
13. Gioe TJ, Maheshwari AV. The all-polyethylene tibial component in primary total knee arthroplasty. J Bone Joint Surg Am 2010;92:478-87.

How to cite this article: Bhende H, Laud N, Deore S, Shashidhar V. Total knee arthroplasty in vascular malformation. Indian J Orthop 2015;49:569-72.

Source of Support: The authors of this study confirm that no financial grant or support from 168 any 169 commercial or noncommercial party was obtained for writing this article, **Conflict of Interest:** None.