CASE REPORT

Treating the 5th Vital Sign: A Case of Bilateral Knee Osteoarthritis in a Patient With Childhood Rickets and Systemic Lupus Erythematosus Undergoing Total Knee Arthroplasty

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ABSTRACT

Deficiencies in Vitamin D effects the growth and development of a children leading to rickets. Presentation of childhood Rickets patients include delayed growth and bowed legs leading to back and arthritis of the knees and hip. We describe a 45-year-old Chinese female with Childhood Rickets and SLE who presented with bilateral knee pain for the past 8 years. This short stature lady had an antalgic gait, marked genu varum on both knees and had limited range of motion of the knees. She undergo total knee arthroplasty for both knee on two different setting. Special consideration unique to this patient population can be discussed such as challenges faced in difficult primary total knee arthroplasty, types of implant to choose to achieve balanced knee, as well as functional problems that may arise post operatively. The patient had a good outcome from her surgery, however this paper alone may not represent all the patient with angular deformity of knees who undergo total knee arthroplasty.

Keywords: Rickets, Total knee arthroplasty, Severe varus deformity

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INTRODUCTION

Rickets is a bone disease that is characterized by deficiency in Vitamin D (calciferol), calcium, or phosphate. Among the causes of rickets are nutritional, gene mutation that leads to dysfunction in protein dependent metabolism, and as a complication from liver or renal abnormality which influences vitamin D metabolism and regulation of mineral in the body. Nutritional rickets are the most common type found globally and according to Malaysian Health and Adolescent Longitudinal Research Team study (MyHeARTs), only 7.4% of adolescents had adequate levels of vitamin D from daily intake. Clinical features of rickets are somewhat heterogenous and the manifestation are age related. Infant may manifest as delayed walker while an adolescent may

present with trivial fracture. Other common clinical features are frontal bossing, delayed teeth eruption, rachitic rosary, and Harrison's sulcus. Radiographic features that are suggestive for rickets are widening of metaphysis, periosteal cupping and the bone may seem osteopenic.

We describe a patient with severe extra – articular deformity of both lower limb with underlying rickets suffering from knee osteoarthritis undergone single stage total knee arthroplasty (TKA).

CASE REPORT

We describe a 45-year-old Chinese female with Childhood Rickets, SLE and dyslipidemia who presented to our clinic with bilateral knee pain for the past 8 years. She was seen in our center throughout the years for pain over the lower back and bilateral knees. She was treated conservatively with pain management but insisted for options of intervention for her severe bilateral knee pain. Denies

having persistent hip or back pain however the knee pain is no longer bearable throughout the last two years requiring daily use of analgesics. Clinically, this short stature lady, walks with an antalgic gait. Marked genu varum can be seen with right knee appears to have more significant deformity than the left. Palpation reveals no bony tenderness and range of motion (ROM) of bilateral knees were limited to 15 degrees to 90 degrees bilaterally. This patient also has pronounced limb length discrepancy of 3 cm seen over left lower limb. Radiologically, her x-ray (Figure 1) showed tri-compartment bilateral knee osteoarthritis Kellgren - Lawrence grade IV, with recognizable bilateral femur and tibia bowing with "Harris - line" noted over her left distal femur. However, her hip joint appears normal.



Figure 1 : Standing x-ray of right and left lower limb with severe varus deformity. Unable to determine the anatomical axis of the femur due to its deformity.

She underwent two separate TKA surgeries from the month of September 2019 and December 2019 for her right knee followed by left. Surgery was conducted using cemented P.F.C Sigma Total Knee System (Depuy®Synthes) without computer navigation. Posterior – stabilized implant (cruciate – substituting) was chosen by the operating surgeon as a preference. Proximal tibia cut was performed prior to distal femur cut and intramedullary guide was used for femur while extramedullary guide was used for tibia. The intramedullary guide was inserted as maximally allowed by patient's femur bowing. Minimal bone cut was performed as well as sequential soft tissue release for a varus knee to achieve balance. The aim of bone cut and soft tissue release was to bring the

mechanical axis of lower limb to a near normal value however not aiming for a perfect restoration. Patella resurfacing was not done as degenerative change was negligible.

Her post-operative x – ray shown improvement of varus deformity but with a mismatched joint line of the knee as this patient had a prior limb length discrepancy (Figure 2). Post-operatively, bilateral knee ROM showing good flexion exceeding 90 degrees with slight lag of extension. Patient's limb length discrepancy was addressed with shoe raise and upon review in clinic four month postoperatively, patient was satisfied with her outcome of surgery as the pain in both her knee was significantly improved.



Figure 2 : Post-operative x-ray of right and left lower limb showing correction of varus deformity.

DISCUSSION

Rickets is a bone disorder associated with abnormal serum calcium and phosphate level and the patient may present heterogeneously depending on the age of onset as well as pathogenesis and feature of short stature, bowing deformity of the leg and widening of the joint(1). The illness once termed as 'The English Disease" was first described in the 1660's resulting in defect of bone mineralization which may lead to angular and rotational deformities of the long bones as well as growth retardation.

Baldini et al. described challenges faced in difficult primary TKA such as patients with many previous incisions, severe coronal deformity, genu recurvatum, stiff knee, extra – articular deformity, post osteotomy of femur or tibia, and neglected patella dislocation. Severe coronal deformity of the knee is those with fixed

varus or valgus deviation of ≥20° from the mechanical axis on weight-bearing film typically have some bone erosion and/or condylar dysplasia on the concave side of deformity while the convex side is affected by the tension force of soft tissues (2). The use of posterior stabilized TKA as compared to cruciate retaining implant in severe varus deformity reportedly having more superior outcome in term of correction of alignment, better knee flexion, and significant improvement of postoperative knee score. Our patient with congenital rickets does manifest the deformity as described as she had 25° varus deformity on the right knee and 26° varus deformity on her left. The implant chosen was a posterior stabilize TKA and we manage to achieve good postoperative alignment and ROM of the knees.

Functional outcomes following TKA such as limited ROM, continuous pain, as well as instability of the joint may be debilitating. There are six functional outcome abnormalities described by Ulrich et al. such as knee flexion contracture, quadricep muscle weakness, knee flexion deficit, limb length difference, foot and ankle malalignment, and peroneal nerve entrapment (3). Limb length discrepancy are well tolerated by patient who undergone both total knee surgery despite having difference of more than 2 cm and patient tend to notice the difference in length in unilateral TKA as compared to bilateral. We decided for this patient to undergo simultaneous TKA in a relatively short time (3 month) as the concern for her apparent limb length discrepancy which might hinder her overall recovery. Although this patient was advice for orthosis to compensate for her lower limb length differences, she chose not to use the orthosis and felt comfortable without one. Oxford Knee Score was used to evaluate patient's outcome post-surgery. Patient scored 36 for right knee and 34 for left knee. This scoring is based on the revised scoring method by Murray et al in 2007 in which best score is 48 for each operated knee.

Current concept of managing patients with angular deformity of lower limb with concurrent knee joint disorder advocates either two stage or single stage deformity correction with arthroplasty. However, in this patient, we can attain a good functional outcome postoperatively despite performing conventional TKA. Parratte el al. and Abdel et al. conducted a retrospective study in comparing functional outcome of TKA implant in mechanically restored patients versus those who didn't. In these retrospective studies, the authors concluded that mechanically align lower limb post TKA does not provide better functional outcome nor superior implant survival after 15 to 20 years (4,5). However, they were unable to recommend range of extreme mechanical axis for reference. Another reason we choose not to perform corrective osteotomy of the femur as there is concern

regarding union and remodeling process especially in this group of patients (2).

Our patient in this case report represents a very selective group of patients as compared to the other typical knee osteoarthritis patients. The total knee surgery performed was aimed to alleviate pain as well as improve ROM of knee and not to gain normal mechanical axis of the lower limb. Through literature search, currently there are no standard guideline as how to approach this group of patients. We performed single stage TKA for her by considering factors that are involve in her disease progression. In the new era of medicine, computer navigated surgery as well as patient specific implant might be considered to cater this type of patients although it is not widely available. Further studies required to better understand and evaluate patients with underlying rickets for improvement of management.

CONCLUSION

We are able to demonstrate, even with severe angular deformity of the lower limb, primary conventional TKA is still feasible. Angular deformity correction of a severe varus or valgus deformed knee pose a challenge for an orthopedic surgeon. It requires fundamental understanding of surgical technique as well as implant design to tackle the intricate manner of the knee defect. Patient's expectation and outcome need to be carefully evaluated to achieve successful surgery and functional painless joint.

REFERENCES

- 1. Carpenter TO, Shaw NJ, Portale AA, Ward LM, Abrams SA, Pettifor JM. Rickets. Nature Reviews Disease Primers. 2017;3.
- 2. A. Baldini, L. Castellani, F. Traverso, A. Balatri, G. Balato VF. The difficult primary total knee arthroplasty. Bone Joint J. 2015;183–96.
- 3. Ulrich S, Bhave A, Marker D, Seyler T, Mont M. Focused rehabilitation treatment of poorly functioning total knee arthroplasties. Clinical Orthopaedics and Related Research. 2007;(464):138–45.
- 4. Abdel MP, Ollivier M, Parratte S, Trousdale RT, Berry DJ, Pagnano MW. Effect of postoperative mechanical axis alignment on survival and functional outcomes of modern total knee arthroplasties with cement: A concise follow-up at 20 years. Journal of Bone and Joint Surgery American Volume. 2018;100(6):472–8.
- 5. Parratte S, Pagnano MW, Trousdale RT, Berry DJ. Effect of postoperative mechanical axis alignment on the fifteen-year survival of modern, cemented total knee replacements. Journal of Bone and Joint Surgery Series A. 2010;92(12):2143–9.