

A rare case of popliteal pterygium syndrome

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This study is performed in Department of Orthopedics, GRMC, Gwalior

Abstract

Popliteal Pterygium Syndrome is an extremely rare genetic disorder which can present with multiple body anomalies, especially the musculoskeletal anomalies.

The very striking characteristic is presence of popliteal pterygium contracture; this connective tissue band can extend from ischial tuberosity to the calcaneum which can severely restrict range of motion, knee extension, abduction and rotation. This case report presents a case of 3-year-old girl child with significant unilateral deformity in left lower limb. Examination revealed extensive popliteal web/contracture left side with associated deformity at left ankle. Serial surgical correction was done with uniplanar ex fix (Distractor) application followed by Tendo Achillis tenotomy and serial splinting with regular follow-ups done. The child was treated successfully with satisfactory results

Keyword: popliteal pterygium syndrome, rare genetic disorder, musculoskeletal anomalies

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How to cite this article

Hans A, Bajoria R S, Singhal V. A rare case of popliteal pterygium syndrome. Ortho J MPC. 2023; 29 (2):62-63
Available from:
<https://ojmpc.com/index.php/ojmpc/article/view/180>



Introduction

Popliteal pterygium syndrome is a rare cause of congenital knee flexion in children associated with joint stiffness and retraction of the posterior soft tissues with a popliteal scar contracture posing therapeutic difficulties. Hereby reporting a case of 3 years old child of popliteal pterygium syndrome treated surgically with 6 months of follow up.

Case report

A 3-year-old girl admitted to orthopaedics department with complain of congenital left lower limb deformity; diagnosed with Congenital Popliteal Pterygium without significant family history of Popliteal Pterygium Syndrome. Child had normal general physical examination. Clinical examination revealed extensive left sided popliteal web extending from ischial tuberosity to calcaneum. Webbing prevented extension at the left knee with the flexion angle of 100 degrees of flexion with equinovarus deformity in left foot and contralateral adaptive knee flexion.

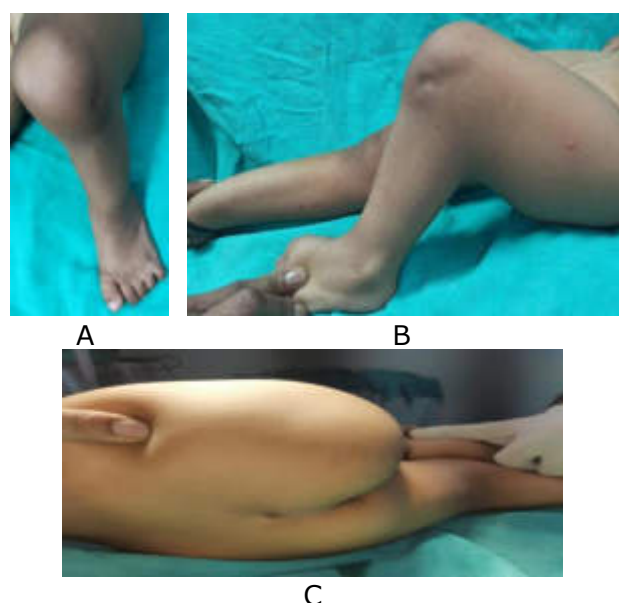


Figure 1- A, B & C- Clinical picture

Spinal pit was also observed. Uniplanar external fixator (distractor) was applied for 3 months with 1 mm (0.5 daytime and 0.5 mm nighttime) daily distraction. After 3 months distractor was removed and Tendo Achillis

tenotomy was done for equinus correction followed by corrective cast application.

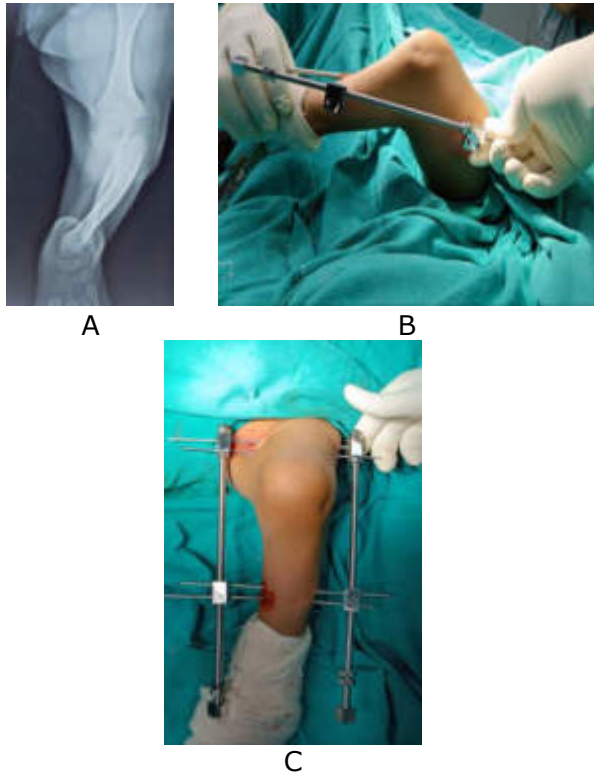


Figure 2- A, B & C Pre op Xray and intra op picture

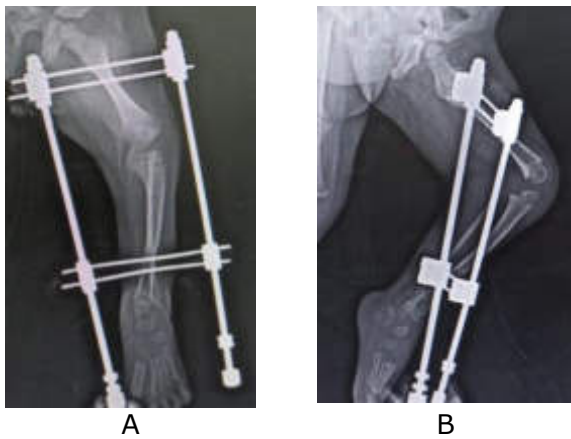


Figure 3- A & B – Post op x-ray

Result

Almost complete flexion deformity correction was achieved with remaining 10 degrees of flexion deformity. Left sided significant shortening was present. Patient is on regular physiotherapy for remaining correction. Further improvement is still expected with time, with regular splinting and follow up.

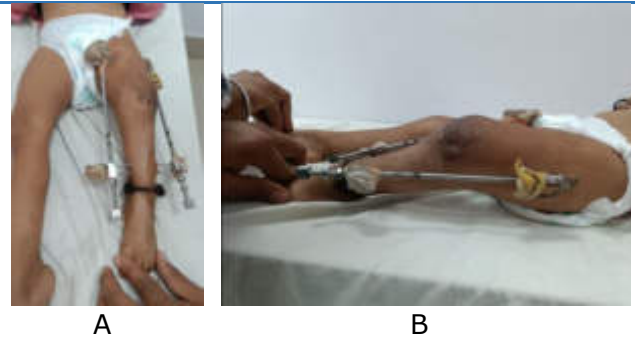


Figure 4- A & B, 6 weeks follow up

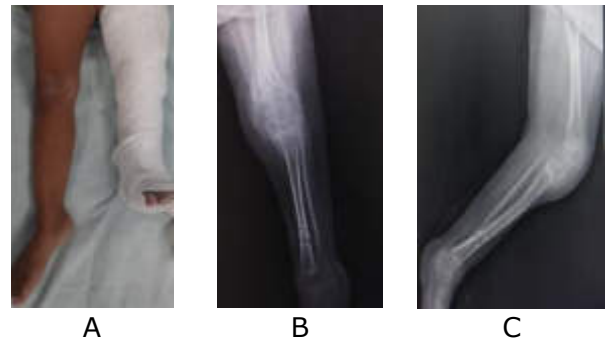


Figure 5- A, B & C, 12 week follow up after implant removal

Conclusion

Popliteal Pterygium Syndrome is a rare genetic disorder, inherited as an autosomal dominant trait. The correction of fixed flexion of the knees in patients with PPS is very difficult. A Doppler Ultrasound and MRI can help formulating the preoperative plan, based on exact neurovascular position. Early Surgical correction is advisable.

References

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