Pterygium Popliteal Syndrome concerning a Case in the Pediatric Surgery Department of the Donka National Hospital (Conakry CHU)

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Introduction

Popliteal pterygium syndrome is a rare birth defect, combining craniofacial, genitourinary and musculoskeletal abnormalities. It is an autosomal dominant disease caused by a mutation in the IRF6 gene. We report in this observation the 1st Guinean case corrected by the surgical method as well as a review of the literature for a diagnostic and therapeutic approach.

Patient and observation

We present the case of a 7 day old male newborn weighing 2700g who was received for bilateral cleft lip and palate, lower lip fossa or sinuses, bilateral popliteal pterygium, and triangular skin fold. above the hallux. The patient underwent several surgical procedures aimed at correcting these abnormalities. The correction of the pterygium of the lower limbs was ensured by excision of the fibrous band, the tenoplasty in z of the calcaneal tendon on the right side and the skin plasty in z in series then immobilized by plaster splints. The immediate postoperative followup was straightforward.

Conclusion

Popliteal pterygium syndrome is a rare congenital malformation, the diagnosis is primarily clinical. Early soft tissue lengthening surgery and serial z-skin plasty provide better correction of the knee pterygium. Correct correction of facial abnormalities gives the child a better appearance The management of this syndrome is multidisciplinary.