

Images in Clinical Practice

Popliteal Pterygium Syndrome

A one-month-old female child weighing 2.7 kg presented with bilateral web over the popliteal region (*Fig. 1*). The clitoris was hypertrophied and a single urogenital opening was present. The facies were normal. A clinical diagnosis of Popliteal Pterygium syndrome was made. An abdominal ultrasound revealed small displaced kidneys, small bladder, and uterus could not be visualized. The cranial ultrasound was normal.

Popliteal Pterygium syndrome is characterized by the presence of web, which may extend from heel to ischial tuberosity. There is also associated short sciatic nerve and other fascial and genitourinary findings because of which it is also known as Fascio-

Genito-Popliteal syndrome. So far no one has reported in literature the exact incidence of this syndrome. It is said to be autosomal dominant with mutation of the IRF 6 gene (Interferon regulatory factor 6).

Clinically, it is defined as an inherited condition with a web beneath the knee (a pterygium is a wing like triangular membrane). The syndrome goes by a number of names including Popliteal web syndrome, besides the Fascio-genito-popliteal syndrome. Presence of web is diagnostic feature of this entity having associated findings in the form of fascial malformation, genitourinary malformation (as in this case). It is to be differentiated from Van Du Woude syndrome, which also has web like malformation but has different etiology.



Fig. 1. Popliteal web in both lower limbs and hypertrophied clitoris.

Various modalities have been tried in the treatment of this condition. The treatment of web is difficult because of presence of short sciatic nerve in the free edge of the web. For extension of the knee, surgery resection of the fibrous bands, freeing of sciatic nerve, Z-lengthening of the Achilles tendon and multiple Z-plasties have been done with successful results.

**Mukesh Gupta,
Rakesh Jora,**
*Department of Pediatrics,
Regional Institute of Maternal & Child
Health,
Umaid Hospital for Women & Children,
Dr. S.N. Medical College,
Jodhpur (Rajasthan) 34200,
India.*

