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Images

An uncommon case of upper limb pterygia

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ABSTRACT

Reporting images in a case of a 9-year-old boy who presented with bilateral congenital webbing (pterygium) of axillae and elbows. This deformity was restricting his axilla and elbow movements. This was successfully treated on one side with multiple Z-plasty. An outline of multiple pterygium syndrome is given herewith.

KEY WORDS

Upper limb webbing pterygium, pterygium, Z plasty

CASE REPORT

A 9-year-old boy presented to the outpatient department with bilateral webbing from the lateral chest wall extending up to his elbows, which looked like wings [Figure 1]. This deformity had been present since birth and involved anterior axillary fold as well as medial elbow [Figure 2]. It was restricting his shoulder and elbow movements. He had no similar webbing anywhere else in the body. There was no history of consanguinity or similar deformity in the family. This was successfully treated on the left side with multiple Z-plasty operation (parents consented for only one side initially).During the procedure, the fibrous bands were excised down to deep fascia and Z-plasty flaps were transposed to lengthen the axillary and elbow contractures [Figures 3 and 4].

DISCUSSION

Upper limb pterygia is described but is uncommon. It can be a part of multiple pterygium syndrome,[1] also referred to as Escobar syndrome. It is a rare autosomal recessive disorder, which is characterized by a web across every flexion crease in the extremities, most notably the popliteal space. In addition, this syndrome is associated with other structural anomalies: a vertical talus, vertebral segmentation anomalies and congenital lordoscoliosis. Other associated anomalies are multiple joint webs, unusual finger contractures, syndactyly, rocker bottom feet, ptosis, antimongoloid slant of palpebral fissures, epicanthal folds, cleft lip and or palate, highly arched palate, scoliosis and short stature.[2]

Autosomal recessive inheritance of multiple pterygium syndrome was suggested by findings in affected siblings with normal parents. It is differentiated from other types of inheritance like autosomal dominant type of multiple pterygium syndrome, autosomal dominant type of popliteal pterygium syndrome and X-linked dominant inheritance variety. Pterygia seen in this variety of multiple pterygium syndrome of neck, antecubital and axillary areas are not

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Table 1: Showing improvement of left upper limb function post-operatively

	Pre op.	*Post op.*	*Improvement*
Shoulder extension	35	50	15
Shoulder abduction	65	90	25
Elbow extension	145–130	145–0	15

Figure 1: Bilateral Upper limb webbing (Pterygia)

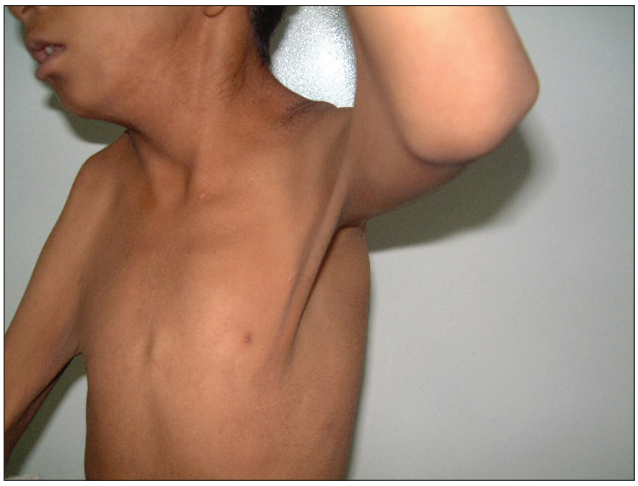

Figure 2: Webbing involving only anterior axillary fold

Figure 3: Multiple Z plasty

Figure 4: Early post op. result

present in the popliteal pterygium syndrome. Cleft lip (with or without cleft palate) has been seen in nearly all cases of popliteal pterygium syndrome, whereas cleft palate is present in only 41% of multiple pterygium syndrome.[3]

Intellect is usually normal in these cases, although there is a danger of mental retardation being assumed due to the multiple congenital defects, misdiagnosis, and delayed motor milestones. Many patients maintain a surprising degree of mobility despite their disabilities. Manual dexterity is often better than predicted from the appearance of the hands[4] as seen in our case.

Multiple pterygium syndrome is uncommon and may present with varying degree of deformities. Correction of restricting webbing by simple Z-plasty adds to the mobility of the affected limb. A joint approach with a team of paediatrician, genetics consultant and orthopaedic and plastic surgeon would be beneficial for a successful outcome.

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