Correction of Webbed-Neck Deformity in Turner’s Syndrome

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Summary. A case of Turner’s syndrome with the typical marked webbing of the neck is presented. A posterior approach is preferred for correction of this webbed neck deformity. A butterfly shaped portion of excess skin is excised and the margins of the defect mobilised as four flaps. The flaps are sutured with a very short midline vertical component, resulting in an almost X-shaped scar. The first procedure resulted in only a partial correction of the deformity and an excess of hypertrophic scar tissue. This was treated with a repeat of the procedure but the midline wounds was repaired with multiple small Z-plasties. The final appearance of the neck was satisfactory in the lateral profile, but there was still some excess tissue posteriorly.

Key words: Turner’s syndrome – Webbed-neck deformity – Lateral and posterior approach.

In 1938 Turner [13] described a syndrome comprising the triad of infantilism, congenital webbed neck and cubitus valgus deformity of the elbow. The webbing of the skin on the lateral aspect of the neck was actually first described by Kobylniki in 1883 [7]. However, it was thought at that time to be a variant of Klippel-Feil syndrome with only the soft tissue abnormality but no bone features. Turner’s syndrome is now recognised as a chromosomal disorder characterised by the presence of a 45 XO configuration in a phenotype female [6]. Further features which are now included in the syndrome are a low nuchal hair level, prominent ears, epicanthal folds, pectus excavatum, micrognathia, high arched palate, middle ear problems resulting in possible deafness, gonadal dysgenesis, renal abnormality and coarctation of the aorta [6, 11].

Sharpey-Schafer, in 1941 [11], demonstrated on post mortem findings that the web of the neck consisted simply of a fold of skin without muscle. This can readily be verified by manually displacing the lax skin of the neck and back of the scalp posteriorly to obliterate the webbing and display
Fig. 1 a–c. Anterior, posterior and lateral views of webbed-neck deformity of Turner's syndrome and the marked pectus excavatum deformity of the case.