

Surgical Correction of Pterygiumcolli: Management of Two Cases and Literature Review.

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Abstract:

The palmar neck is a very rarely described in the medical literature. It is usually associated with Ulrrich-Turner Syndrome (a chromosomal disease related to the complete or partial absence of an X chromosome). Several surgical techniques can be used to treat this malformation, but in severe forms it is hard to completely correct the palm and the low hair implantation.

Through two clinical cases observations, the authors highlight two simple, reliable and reproducible surgical techniques based on a double Z-plasty in one patient and a single asymmetric Z-plasty described by Servant in the other one. Achieving a satisfactory result with hidden scars and consequently a significant improvement in neck shape, which has a positive impact on body image and further relationship life.

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I. Introduction :

The webbed neck was first described in 1883 by Kobylinsk [1], while the alternative term "pterygiumcolli" was coined in 1902 by Funke from his Greek explanation pterugion "little wing" and collum "neck" [2]. In 1938, Turner described a series of women with a syndrome of infantilism, an ulna valgus and the presence of a lateral skin fold of the neck, thus making the first association of pterygiumcolli with a clinical syndrome [3]. This syndrome, which has since been named after Turner, has an abnormal karyotype (45X0) that may be present in a variable percentage of cells. Clinical signs include: female phenotype, epicanthus, palmar neck and ulnar valgus. The low hairline implantation that extends to the nape of the neck and palmar neck extends from the mastoid to the acromial region. Pterygiumcolli may also be associated with other syndromes such as Noonan's syndrome and trisomy 13, 18 and 21.

II. Material And Methods:

Observation N°1 :

Our first patient, 9 years-old, who presented with a palmar neck with low hair implantation, was characterized by a thick and fibrous band extending from the mastoid to the acromial region, limiting head rotation movements and the wearing of specific clothes, as well as an aesthetic discomfort with important psychological repercussions on this young pupil (Fig.1). The clinical picture is also associated with a small stature with a large shielded thorax and protruding ears. The karyotype was 46XX/45XO , compatible with Turner syndrome.



Figure 1: Preoperatively clinical appearance of the patient.
Images 1, 2 et 3: palmar appearance. Image 3: Low hair
implantation.

Observation N°2 :

Our second patient is a young lady, 22 years old, single, no schooling education, from a low socio-economical family. She has no pathological history and has a short neck with a palmar which extends from the mastoid to the base of the lateral cervical region. Low hair implantation and poorly hemmed ears. (fig.2) The external genital organs were normal-looking and secondary sexual characteristics were present.



Figure 2: Preoperative clinical aspect of the second patient.
Image1: front view. Image 2: side view

Surgical techniques:

To correct the malformation, we performed a mixed technique on the first patient combining: a double Z-plasty and resection of excess skin.

Our incisions were as far posterior as possible so it could be hidden. Thus, these incisions were located along the posterior end of the vertical part of the band, at the level of the retro auricular groove at the top, one parallel to the latter passing through the base of the nape of the neck at the level of the distal end of the web, and another parallel crossing the middle of the web (fig. 3).



Figure 3: incisions drawing.

Four greasy skin flaps were dissected in a subcutaneous plane in order to isolate and transpose them. The excess skin was resected, essentially in the posterior flaps, which had a very hairy skin, thus reducing the problem of low hair implantation. The platysma was neither thick nor excessive, and played no role in the malformation (Fig.4).



Figure 3: left: flaps raising with intraoperative appearance of the bands, right: flaps transposition.

No suction drain was carried out, however, the upholstery was done with 3/0 resorbable stitches. This increased the adhesion of the flap and also ensured a harmonious continuity between the neck and shoulders. The skin was sutured with monofilament in 2 planes: deep 3/0, and superficial with an intradermal overjet 4/0.

In the second patient our technique consisted of an asymmetric Z-plasty described by Servant. It is a transposition flap in which two internal alternating triangles exchange their adjacent sides.

In our technique, the Z is centered on the skin fold which is partially excised with detachment and release of fibrous adhesions on both sides.

On the posterior side of the neck, the lower branch of the Z follows the hairline, separating from the vertical branch of Z at an obtuse angle.

The other upper branch, on the other hand, separates from the vertical branch of the Z at an acute angle and descends in the carotid region to the anterior face of the fold, thus creating the asymmetrical Z line.

The two alternating triangles thus formed exchange their sides.

The closure is performed on a two-plane suction drain with a compression dressing.

This technique has the advantage of being simple, easy to reproduce and above all is done under local anesthesia. Also, this technique makes it possible to partially treat the almost constant low hair implantation in the pterygium colli syndrome.

III. Results :

There were no complications, and an adequate neck shape was obtained, as well as a lower hair implantation. The patients left the hospital the next day.

Today, the head rotation movements are no longer limited, and our patients can dress as they wish and leave the hospital without any aesthetic concerns (Fig.5).

The functional and especially aesthetic results are considered good by both the surgeon and the patient.



Figure 5: Postoperative appearance of the first patient.



Figure 5: Postoperative appearance of the second patient.

IV. Discussion :

Very rarely described in the medical literature, the surgical correction of pterygium colli has the following objectives: restoration of a normal neck morphology, restoration of normal movements and posterior correction of low hair implantation [3-5].

The etiology is still unknown. Several theories that may explain this phenomenon have been formulated, but remain controversial. The first theory suggests that there is a pathological growth differential between the cervical and acromial regions during the third month, when the neck is too long in relation to the rest of the body and that during the same period, this structure is located on the same vertical plane as the acromion, which predisposes to the development of the disease [3]. A second theory suggests the presence, during embryonic life, of a significant cystic hygroma in the posterior neck region, which during the regression process would leave after-effects in the lateral cervical skin.

There are different techniques and operating procedures for correcting the neck strap. Most of them are adapted around the need to correct an abnormal hair root and bands of subcutaneous tissue. Such as the lateral and posterior advancement of flaps with a long posterior scar but an adequate shape [6], the tissue expansion in two stages, with significant morbidity [7] and finally The "butterfly method" in which through a posterior approach involved a skin resection similar to the shape of a butterfly, the result considered as insufficient, due to the thickness of the tissue strip fibrous with a large scar.

We present two surgical techniques with satisfactory results that resolved the vertical defect, the transversal excess of skin, the removal of thick tissue fibro-adipux and the correction of the low hair implantation. The scar has a slight unsightly lateral extension, but it remains easily concealed.

The techniques include :

A double Z-plasty modified, under general anesthesia for a severe case of pterygium colli.

An asymmetric Z-plasty described by Servant, under local anesthesia, for another patient with a relatively smaller web.

V. Conclusion :

We think that both techniques are useful and adapted for each patient. They have led to a significant improvement in the neck shape of both our patients with a positive impact on body image and relationship life.

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