

ANGIOKERATOMA CIRCUMSCRIPTUM – SUCCESSFUL TREATMENT WITH A NEW TECHNOLOGY

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Summary. The term angiokeratoma means hyperkeratotic, dark-red to purple papules, with dimensions of 1-10 mm in diameter. Histopathologically they are seen as vascular ectasias in the upper dermis surmounted by hyperkeratosis of the epidermis. Several clinical variants of angiokeratomas exist. Some variants are localized on the genitals (Fordyce's angiokeratoma), on the fingers and toes (Mibelli's angiokeratoma) or form confluent plaques (Angiokeratoma circumscriptum). We present a 15-year old female patient with Angiokeratoma circumscriptum of one year duration, which expands in size with time. Interestingly, angiokeratoma circumscriptum appeared on the place of an operational scar. Laser treatment is one of the effective options for removal of localized angiokeratoma. The spectrum of treatment options includes ablative lasers, argon laser and IPL. In our case we achieved an excellent and rapid therapeutic result using a laser apparatus with elōs technology.

Key words: *Angiokeratoma circumscriptum, laser therapy, elōs technology*

INTRODUCTION

Angiokeratomas are a group of vascular ectasias that involve the papillary dermis and may induce acanthosis and hyperkeratosis of the overlying epidermis. The exact mechanism for their development is unknown. Clinically, they appear as one or several dark-red to purple papules, mostly with a verrucous surface [16]. These vascular anomalies can be presented as a wide range of lesions varying in size, depth and location [1]. Several clinical variants of angiokeratomas exist. Angiokeratoma circumscriptum is the least frequent

of all types of angiokeratomas [13]. Here, we report a case of angiokeratoma circumscriptum developed on the chest subsequently to injury and treated successfully with elōs technology.

CASE REPORT

A 15-year old female patient was presented to our department with asymptomatic papules on the left chest. She had undergone an operation for a cyst on the same place (between the left armpit and the left breast) 7 years ago. Since 1 year on the place of the operational scar purple papules and plaques developed. The lesions grew in size and number. Outpatient treatment with topical corticosteroids was performed without effect.

DERMATOLOGICAL STATUS

Examination revealed several clustered, dark-red to purple colored papules and plaques, measuring 1~ 5 mm in diameter and arranged in zosteriform pattern of distribution on the left chest area (Fig.1 A, B).

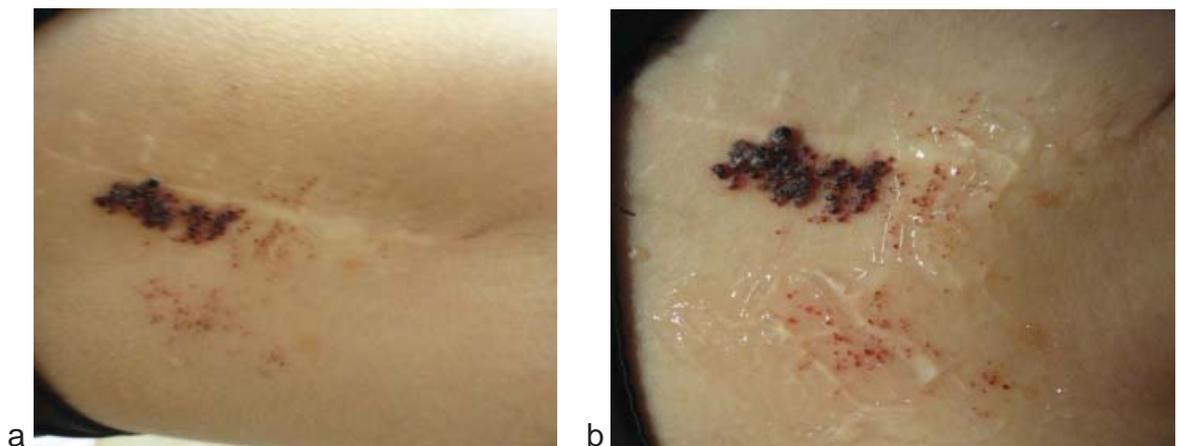


Fig. 1. A, B - Several clustered, dark-red to purple colored papules and plaques, measuring 1~ 5 mm in diameter and arranged in zosteriform pattern of distribution on the left chest area

The diagnosis of after-trauma angiokeratoma circumscriptum was performed (Fig. 2).

We decided for laser treatment, and started the therapy with a laser device using elōs technology (parameters: SRA head using Optical energy 32 J/cm² and Radiofrequency 25 J/cm³). After 2 sessions an excellent and rapid therapeutic result was achieved (Fig.3 A, B). No recurrence has been observed during the follow-up period of 9 months.



Fig. 2. After-trauma angiokeratoma circumscriptum

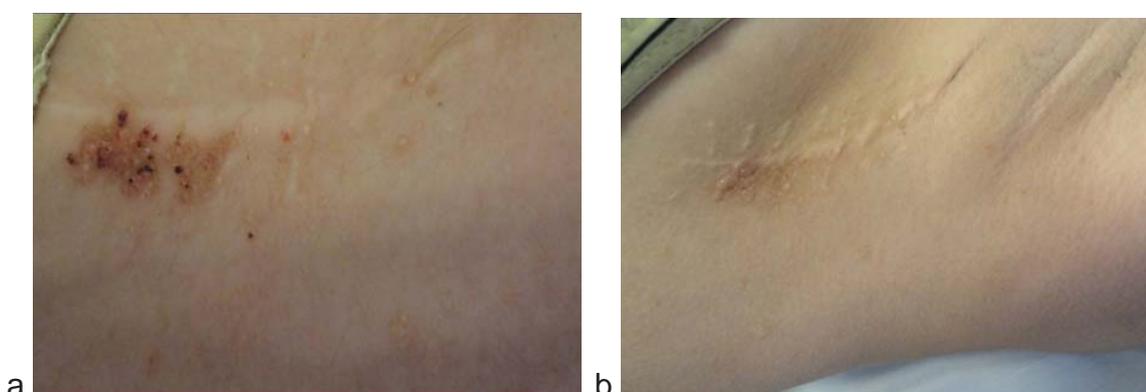


Fig. 3. **A-**Therapeutic result after first procedure; **B-**Therapeutic result after second procedure

DISCUSSION

According to Updated ISSVA classification angiokeratomas are part of the group of the benign vascular malformations. Precisely, angiokeratomas are capillary malformations defined by ectasia of the papillary dermis vessels with secondary epidermal reaction changes, such as acanthosis and/or hyperkeratosis [16].

The mechanism for development of angiokeratomas is unknown. Several causal factors have been proposed: congenital defect, pregnancy, trauma, subcutaneous hematomas, tissue asphyxia [9]. Overall, altered hemodynamics (typically caused by trauma) appear to produce telangiectatic vessels of the papillary dermis with an overlying reactive hyperkeratosis to the epidermis [16, 6].

The histological findings are identical for all types of angiokeratomas and consist of dilated, thin-walled, congested capillaries mainly in the papillary dermis overlying an epidermis that exhibits acanthosis and hyperkeratosis [4]. Elongated rete ridges may partially or completely enclose vascular channels, and a collarette may be present at the margin of the lesions [1].

Many types of angiokeratomas have been described in the literature: angiokeratoma corporis diffusum (Anderson–Fabry disease); solitary or multiple angiokeratoma (Sporadic angiokeratoma); angiokeratoma Fordyce; angiokeratoma Mibelli (Angiokeratoma acroasphycticum digitorum);

angiokeratoma circumscriptum (angiokeratoma corporis naeviform) [7, 15]; angiokeratoma of the tongue (Caviar spots); acral Pseudolymphomatous angiokeratoma of children (Apache) [2].

In 1915 Johannes Fabry described for the first time angiokeratoma circumscriptum as a localized form of angiokeratoma, mostly seen on the skin of the lower limbs or trunk. Angiokeratoma circumscriptum is a capillary-lymphatic malformation which usually develops during infancy or childhood as either a plaque of multiple discrete papules or hyperkeratotic papules and nodules that often become confluent. They occur on the trunk, arms or legs and are unilateral in most patients [1]. Angiokeratoma circumscriptum is quite rare with unknown frequency. Women are affected more commonly than men, in a ratio of approximately 3:1. There is no ethnic predilection.

There has been only one report of angiokeratoma circumscriptum following skin injury [11], which makes our case quite unique. Several reports have noted angiokeratoma circumscriptum appearing on the ventral and, less commonly, the dorsal surface of the tongue [5, 10, 17]. Differential diagnosis could be made with Lymphangioma circumscriptum, Verrucous hemangioma, Unilateral nevoid telangiectasia, Angioma serpiginosa, Pyogenic granuloma, Malignant Melanoma, Blue rubber bleb nevus syndrome, Osler-Weber-Rendu syndrome [8].

The treatment for Angiokeratomas is difficult. Several different methods can be used: surgical excision, laser therapy, cryotherapy, electrocautery (Table 1).

Table 1. Treatment methods – advantages and side-effects

Treatment method	advantages	side-effects
Surgical excision	completely removes the lesions, no recidives	disfiguring scars, long recovery time
Laser therapy	best cosmetic outcome	recurrence, scarring, post treatment hypo- and hyperpigmentations
Cryotherapy	for very superficial lesions	recurrence, scars, many procedures
Electrocautery	for small superficial lesions	recurrence, scars

The treatment of larger lesions usually requires laser therapy, because other treatment modalities may produce extremely disfiguring results. Several laser systems have been used for treatment of angiokeratomas [3, 12, 14]: Argon lasers; Carbon-dioxide lasers; Cooper vapor lasers; Neodymium: yttrium-aluminum garnet (Nd: YAG) lasers; Pulsed dye lasers; Intense pulse light source systems IPL; Elōs laser devices. The elōs technology includes a combination of optical and electrical RF energy (Fig. 4).

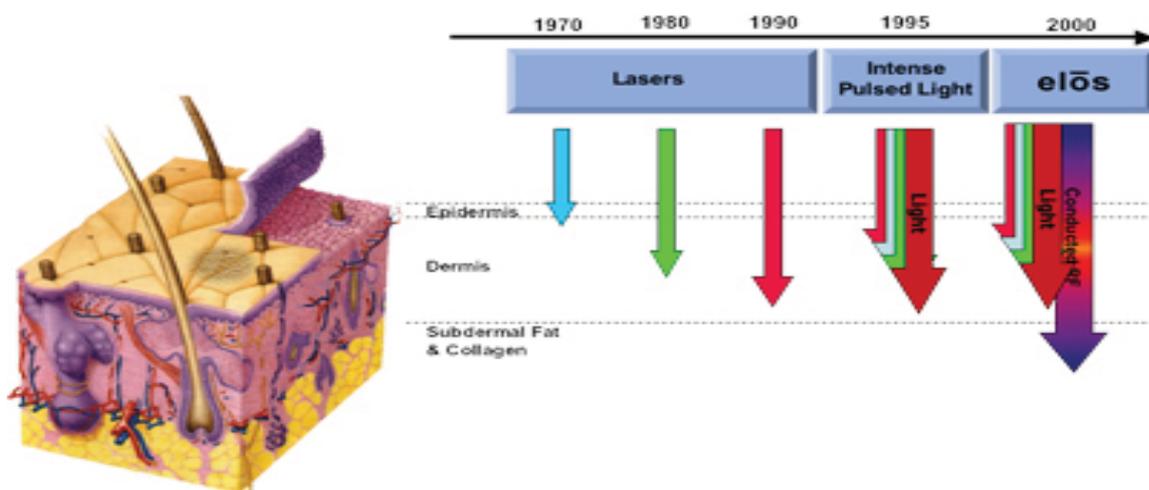


Fig. 4. Elōs technology includes a combination of optical and electrical RF energy

This new technology encompasses a blend of optical and electrical radiofrequency (RF) energies simultaneously applied to the tissue. The rationale behind combining electrical and optical energies is 2-fold: 1) a synergistic effect occurs between the 2 forms of energy and 2) lower levels of both energies can be used, facilitating treatment for individuals of all skin types and hair colors and potentially reducing the risk of side effects associated with either optical or RF energy alone.

According to our search in the literatures, we didn't found any other published reports about angiokeratoma circumscriptum treated with elōs laser technology, so this is the first case of successful treatment with this device. Along with the excellent therapeutic result we can recommend this method as a standard treatment for angiokeratomas (Fig. 5).



Fig. 5. After-trauma angiokeratoma circumscriptum treated successfully with elōs laser technology

CONCLUSION

In conclusion we have presented a rare case of angiokeratoma circumscriptum developed after excision of a cyst on the left chest seven years ago, and treated successfully with the modern elōs laser technology.

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