

Statement of Purpose

Angiokeratoma Circumscriptum is a rare vascular lesion that usually presents in a unilateral band like appearance. Although these lesions are benign and do not require treatment, it is important to recognize them as they can mimic malignant melanoma. Presented below is a review of the literature and case report that highlights this rare lesion.

Introduction

Angiokeratomas are a group of vascular lesions which involve the papillary dermis. They have a prevalence of 0.16% and there are currently 8 sub types described in the literature. Angiokeratoma Circumscriptum is the rarest of these lesions with only 100 reported cases noted in the literature. Usually, these lesions are seen at birth but they can arise in the 2nd and 3rd decade of life. They present as dark red to purplish hyperkeratotic papules and plaques that can have a warty like appearance. Biopsy is needed to rule out other entities that can resemble these lesions. Histopathological exam reveals involvement of the papillary dermis with varying features including ectatic, thin-walled vascular spaces, acanthosis, hyperkeratosis, papillomatosis and dilated capillaries^{1,4}.

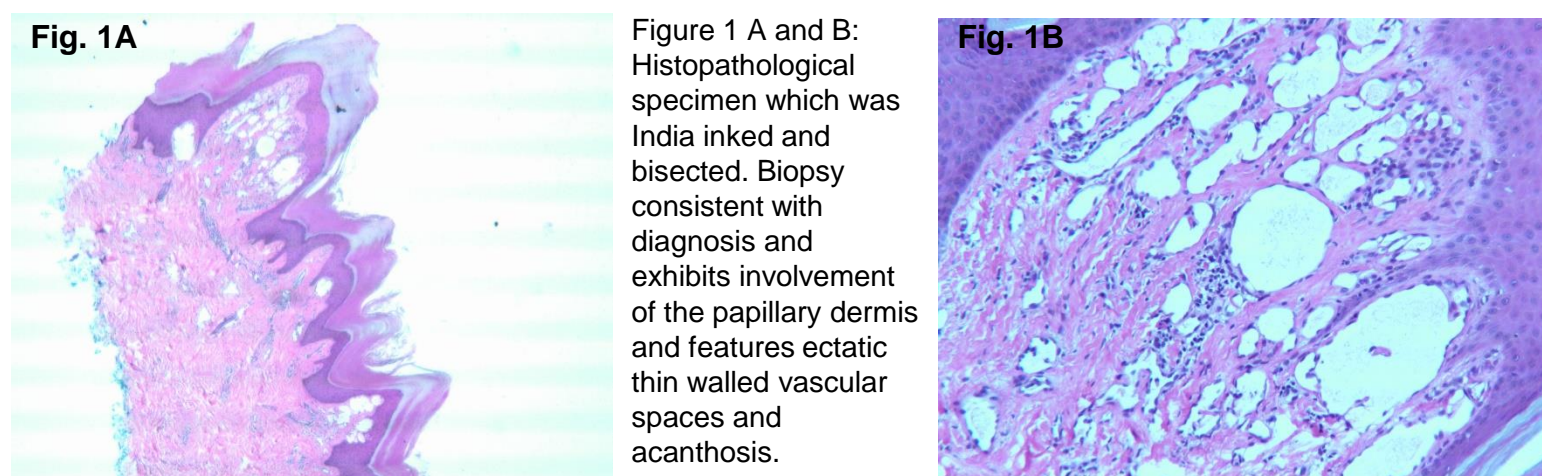


Figure 1 A and B: Histopathological specimen which was India inked and bisected. Biopsy consistent with diagnosis and exhibits involvement of the papillary dermis and features ectatic thin walled vascular spaces and acanthosis.

Differential diagnosis includes verrucous hemangiomas, verruca, nevus, and angioma serpiginosum. Malignant melanoma should also be included in the differential if thrombosis occurs within the lesion which can only be excluded by biopsy⁴. Treatment is not necessary but can be performed if they bother the patient or for cosmetic reasons. Treatment consists of either local excision and electrocautery, cryotherapy or laser treatment. Pozo et al treated two patients successfully with carbon-dioxide laser⁵. Each patient underwent at least four treatment sessions and at the 6 month follow up had no recurrence. Pasyk et al successfully treated two patients with argon laser with an average follow up three years⁶. Presented below is a case report illustrating this rare vascular lesion.

Case Report

A 35-year-old female presented with bilateral pedal skin changes which have insidiously occurred and progressed in size for more than one year. Previous treatment had included consistent use of urea 40% cream with no improvement. The patient's medical history included hypertension and seasonal allergies. On physical examination, the patient's neurovascular status was intact, including the presence of protective touch-pressure sensation. Examination of the dorsal skin to the bilateral digits revealed raised white fibril proliferations with a verruciformis appearance in a patchy distribution (Fig. 2A-B). There were no open lesions, drainage, or signs of infection noted. A biopsy of the skin was taken and histopathological diagnosis of angiokeratoma circumscriptum was made (Figure 1A-B). Conservative treatment was exhausted using treatments of canthacure and topical steroids without improvement. Due to failed conservative treatment, the patient opted to pursue surgical options.



Fig. 2A

Fig. 2B

Fig. 2C

Fig. 2D

Case Report Continued



Fig. 3A

Fig. 3B

Fig. 3D

Fig. 3E

Fig. 3C

Fig. 2A-D: Initial clinical pictures showing multiple white plaques in verruciformis pattern involving the digits bilaterally.
Fig. 3A-E: Intra-operative pictures demonstrating debridement with a 15 blade and subsequent electrocautery of the lesions.
Fig. 4A-B: Three month follow-up showing improvement of the lesions

After adequate sedation, and local digital block, a 15 blade was used to debride excessive hyperkeratotic tissue overlying the lesions (Fig. 3A). Next, using a combination of a spatula blade and needle blade, electrocautery was performed on the lesions. (Fig. 3C). A straight curette was used to remove the resultant eschar (Fig. 3B). The dermis beneath the eschar was noted to be free of further papillary

Case Report Continued



Fig. 4A

Fig. 4B

lesions. The areas of cauterization were covered with a thick coat of silver sulfadiazine followed by a dry, sterile dressing. A three month follow-up of the patient status post surgery revealed improvement in her lesions (Fig. 4A-B). Patient did develop areas of hypertrophic scar but admitted to a history of hypertrophic scar formation. The patient continues to apply hypercare 20% solution once daily before bed to her lesions. The patient's lesions although improved have never fully resolved and the patient may pursue a second electrocautery treatment if continued conservative care does not improve the lesions.

Conclusion

Presented above was a review of the literature and a case presentation depicting this rare vascular lesion. Angiokeratoma circumscriptum is a benign lesion, however a biopsy should routinely be performed to rule out more invasive entities. Treatment recommendations are mostly anecdotal due to the rarity of the lesion, and more studies need to be performed to determine the best course of treatment.

References

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