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Pseudo ainhum and facial malformation secondary to Streeter's dysplasia

Dear Editor,

A 5-year-old girl presented with constricting bands around the first and the fifth fingers of right hand (Fig. 1a-c) appeared since birth and stable over time. Patient history revealed surgical correction of complete cleft lip and palate. Doppler ultrasound of the affected digits did not show any sign of significant vascular constriction and consequent tissue ischaemic suffering. On the basis of these findings, a diagnosis of pseudo ainhum secondary to amniotic bands syndrome was made.

The classical ainhum, also known as dactylolysis spontanea, is an idiopathic and progressive form of constricting bands of the extremities leading in most severe cases to auto-amputation without bleeding. This condition of unknown aetiology usually involves the fifth toe,[1] but the fingers may be rarely affected.[2,3]

Differential diagnosis may be made with pseudo ainhum that refers to constriction bands secondary to congenital disorders such as amniotic bands, palmoplantar keratoderma or acquired condition due to psoriasis, scleroderma and tourquinet.

The amniotic band syndrome, namely Streeter's dysplasia (SD), is a complex clinical condition characterized by a wide severity range of clinical presentation of constricting bands that can potentially affect each part of the body (upper and lower extremities, but rarely the trunk). It is a rare syndrome described for the first time by George Streeter in 1930,[4] with an incidence of 1–10 000–15 000, without sex predilection.[5] The pathogenesis is unclear but the most valuable hypothesis would attribute the formation of bands to the rupture of amniotic membrane. SD is

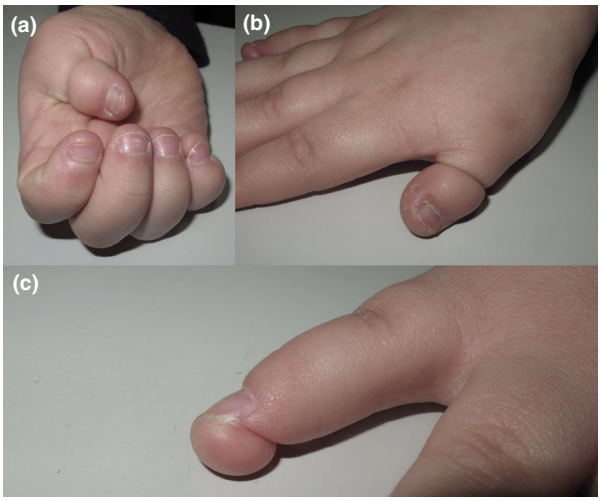

Figure 1 (a) General view of the right hand with constricting bands affecting in particular the first (b) and the fifth (c) fingertips.

associated with other anomalies that include clubfeet, renal and cardiac abnormalities, tibial pseudo arthrosis or facial malformation.[5,6] The extension of facial alteration depends on the time of onset and is characterized by multiple combination of facial cleft, cleft palate and lip, ocular lesions, and cranial malformations. The annular rings occur on various severity degree ranging from superficial, involving cutis and subcutis to deeper involving the deep fascia. Doppler ultrasound is necessary to define the presence of vascular constrictions that may represent the cause of ischaemic suffering and finger/toe auto-amputation. The diagnosis is clinical, although other diagnostic instruments such as radiologic imaging allow to evaluate the tissue involvement degree, especially for deeper annular ring rarely extending to the bone. Currently, the improvement in prenatal diagnosis could allow the diagnosis in utero showing swelling of digits or limbs after the constriction.[7] The treatment is frequently plastic surgery to correct the facial malformation and, in case of necessity, pseudo ainhum decompression to avoid amputation. This latter requires Z-plasty in advanced phase.[8]

It is uncommon to see pseudo ainhum in Dermatology departments and for that reason we were intentioned with our paper to underline the possible scenarios that could be associated with constriction bands trying to give a quick guide to recognize and distinguish each condition, in order to prevent and promptly treat the most feared complication, that is, auto-amputation.

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Dermoscopic features of benign vascular lesions presenting on volar skin: a case series and literature review

Editor

Dermoscopy has allowed for improved diagnostic accuracy of pigmented lesions located on volar skin.[1] However, the dermoscopic patterns associated with benign and malignant amelanotic lesions located on the palms and soles [volar lesions (VL)] are not well established. Herein, we describe a recurrent benign dermoscopic pattern visualized in VL located on either the palms or soles. The recognition of benign patterns manifested by these lesions can help to exclude amelanotic acral lentiginous melanoma from the differential diagnosis, helping to better select which patients should undergo a biopsy.[2]

This was an IRB-approved study. In two dermatologic clinics at New York, NY and Thessaloniki, Greece, 11 pink amelanotic VLs (seven and four, respectively) were identified. Ten of these lesions were clinically and dermoscopically diagnosed as benign VL while one lesion was biopsied. Histopathologic examination of the biopsied lesion confirmed the diagnosis of an angioma. In addition, a literature review using keywords 'angioma', 'hemangioma', 'vascular malformation', 'capillary malformation', 'port wine', AND 'acral' AND 'dermoscopy' revealed a total of three cases, including two cases of angioma serpiginosum.[3–5] All of the lesions included had good quality dermoscopic images, and these were examined by the authors who commented on size, dermoscopic structures, colours and location of dermoscopic structures and their relation with the dermatoglyphics (ridges and/or furrows).

All lesions (n = 14) were located on the palms or soles and none crossed Wallace's line. The size of the VLs ranged from 2 to 12 mm. Eight of 14 lesions (57.1%) revealed only one dermoscopic structure, namely red dots/globules. In all of these eight lesions, the red dots/globules were arranged in a unique distribution, with the dots aligned linearly along each side of the ridge, sparing the eccrine openings (Fig. 1). This unique distribution leads to a 'double red-dotted parallel ridge pattern'.[2] Two of 14 lesions (14.3%) had areas suggestive of the same pattern, but the small size (≤2 mm) made it difficult to conclusively determine the dermoscopic pattern. Two of 14 lesions (14.3%) revealed lacunae, similar to those seen in angiomas on non-glabrous skin. The lacunae in these lesions were aligned along the ridges with some encompassing the entire ridge; however, none crossed over to involve the furrows (Fig. 2). One lesion contained both the double red-dotted parallel ridge pattern and few regular appearing lacunae encompassing the ridges. One of 14 lesions was a