(surgeons, gynaecologists, dermatologists and general practitioners) may see the condition, and where there is often a tendency to avoid invasive and expensive diagnostic procedures.[3] Unlike the classic, frog spawn appearance of lymphangioma circumscriptum, lymphangiectasias of the vulva present as large clusters of vesicles and papules or oedematous polypoid nodules mimicking genital warts.[6] Hence, vulvar LC needs to be differentiated from various dermatological disorders for which treatment methods are different, such as genital warts, herpes zoster, molluscum contagiosum and leiomyoma.[7] Histologically, numerous dilated lymphatics in the superficial and papillary dermis are seen. There is clear fluid and, less frequently, red blood cells in their lumina. In the overlying epidermis there is some degree of acanthosis and hyperkeratosis. The surrounding stroma shows scattered lymphocytes.[8] Recognition and appropriate treatment of vulval lymphangiectasia is important primarily because the lesions may act as portals of entry for infection. In addition, persistent leakage of lymphatic fluid may be mistaken for urinary incontinence.[3] There is no standard therapy for the management of LC. The most common procedures are abrasive therapy, sclerotherapy, electrocoagulation and surgical resection. Even with the best treatment option, recurrence is common.[9] In our case, the obstruction of the lymphatics by an overtly distended gravid uterus appears to be the most plausible explanation for the vulvar lymphangiectasia occurring in the setting of a twin pregnancy, resolving spontaneously in the postpartum period with the physiological involution of the uterus.

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We report a case of lymphangiectasia of the vulva occurring in the setting of a twin pregnancy to emphasize the importance of recognising this condition in pregnancy and differentiating it from other vulvar dermatoses which can present similarly, but may require a different approach to management

References

1 Harwood CA, Mortimer PS. Acquired vulval lymphangiomata mimicking genital warts. *Br J Dermatol* 1993; **129**: 334–336.
2 Mortimer PS. Disorders of lymphatic vessels. In Burns T, Breathnach T, Cox N, Griffiths C, eds. Rook's Textbook of Dermatology, Vol. 3, 8th edn. Blackwell Science Ltd, Oxford, 2010: 48.25–48.28.
3 Verma SB. Lymphangiectasias of the skin: victims of confusing nomenclature. *Clin Exp Dermatol* 2009; **34**(5): 566–569.
4 Mallett RB, Curley RK, Mortimer PS. Acquired lymphangiomata: report of four cases and a discussion of the pathogenesis. *Br J Dermatol* 1992; **126**: 380–382.
5 Verma S. Pregnancy-induced lymphangiectasias of the vulva. *Int J STD AIDS* 2008; **19**(3): 211–212.
6 Peachey RDG, Lim CC, Whimster IW. Lymphangioma of the skin. *Br J Dermatol* 1970; **83**: 519.
7 Vlastos AT, Malpica A, Follen M. Lymphangioma circumscriptum of the vulva: a review of the literature. *Obstet Gynecol* 2003; **101**: 946–954.
8 Mu XC, Tran TA, Dupree M, Carlson JA. Acquired vulvar lymphangioma mimicking genital warts. A case report and review of the literature. *J Cutan Pathol* 1999; **26**(3): 150–154.
9 Roy KK, Agarwal R, Agarwal S, Kumar S, Malhotra N, Gopendru N. Recurrent vulvar lymphangioma circumscriptum - a case report and literature review. *Int J Gynecol Cancer* 2006; **16**: 930–934.

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Colour Doppler ultrasound findings in the nail in cystic fibrosis

Editor

Cystic fibrosis (CF) is an autosomal recessive disease resulting from mutations in the CF transmembrane conductance regulator gene. The main clinical manifestations are related to lung disease, pancreatic insufficiency and infertility.[1] CF can also present with skin changes, however, such cutaneous manifestations seem to be underestimated.[2]

A common, but not specific sign of CF is digital clubbing. Nevertheless, the pathophysiology of clubbing still remains unclear. Possible causes of clubbing have included several factors such as hypoxia, platelet activation, releasing of platelet-derived growth factor and vascular endothelial growth factor.[3,4] Prostaglandins, ferritin, bradykinin and oestrogen have also been implicated in the genesis of this morphological alteration.[5]

The 'spongy' feel of the nail have been postulated to be caused by a fibrovascular hyperplasia of the underlying connective tissue of the nail bed.[6] A high presence of electrolytes such as sodium and chloride has been reported in the nail bed of the CF patients.[7]

Ultrasound has been proven as a useful imaging technique for studying the nails non- invasively. Thus, a wide range of inflammatory and tumoural conditions have been well described on sonography.[8,9] To date, there are no reports on the imaging characteristics of the nail bed in CF.

We report the case of a 25-year-old male CF patient who was diagnosed by the age of 7. The physical examination revealed clubbing in both hands (Fig. 1). To rule out underlying nail pathology an ultrasound examination of the ungual region was requested.

The sonographic examination demonstrated a diffuse increase of the thickness and decreased echogenicity of the nail bed in the fingernails of both hands which was more important on both thumbs. This alteration involved the matrix region in all nails and was accompanied by an upward displacement of the ungual plates. The proximal part of the ungual plates in both thumbs was irregular being the latter finding more prominent in the left thumb. In some of the fingers loss of the proximal part of the

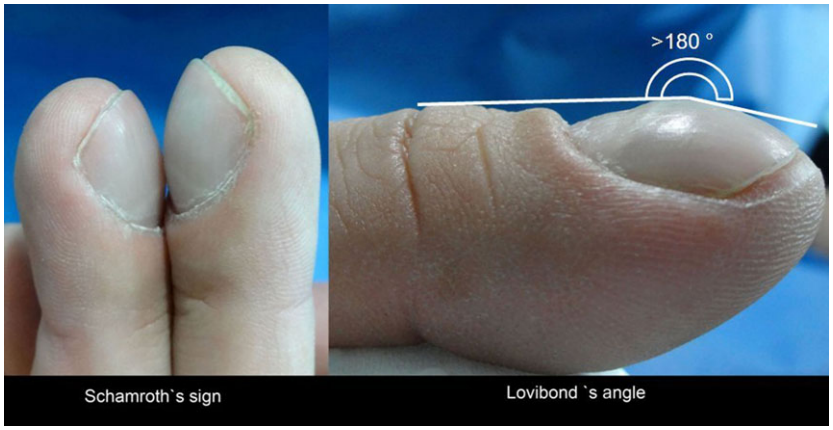

Figure 1 Clinical photographs of the case show positive Schamroth`s sign and Lovibond`s angle.

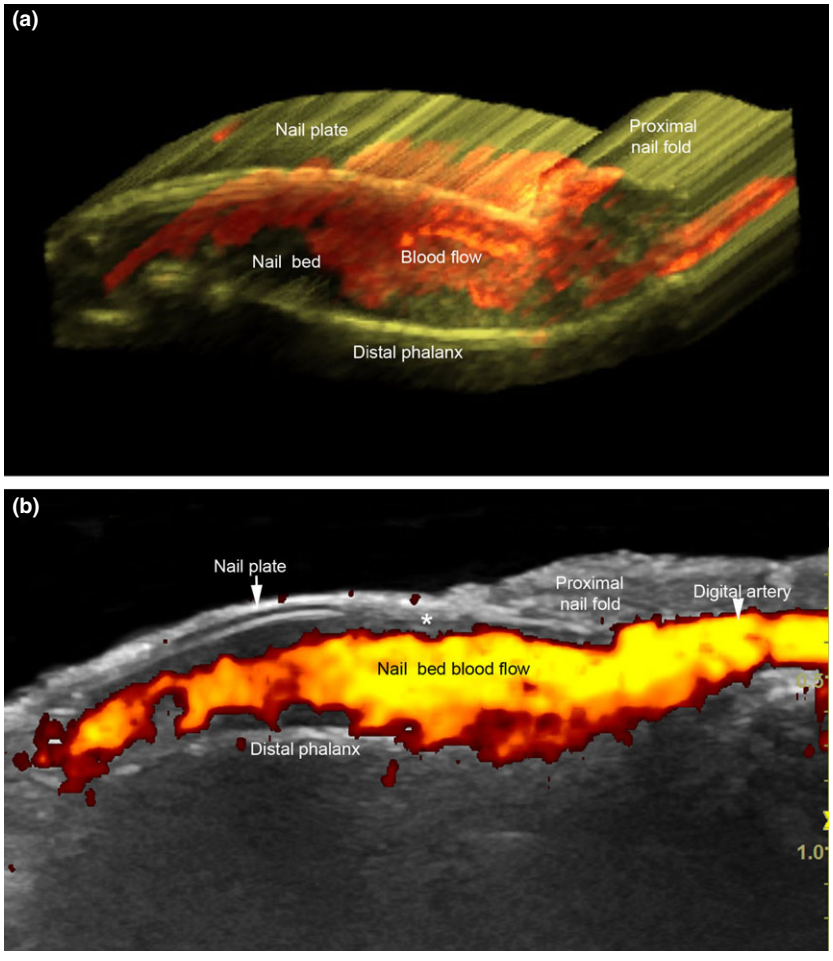

Figure 2 (a) 3D power angio reconstruction demonstrates increased thickness and blood flow in the nail bed of the right thumb. Notice the upward displacement of the nail plate. (b) Power Doppler ultrasound image of the right ring finger demonstrates prominent hypervascularity within the nail bed and a dilated digital artery. There is upward displacement of the nail plate and loss of definition (*) of the proximal part of the ventral plate.

ventral plate was noticed. No solid or cystic nodules were detected within the nail bed. The colour Doppler ultrasound with spectral curve analysis showed hypervascularity in all the fingernails of both hands, mainly affecting the proximal part of the nail beds. There were abundant arterial vessels in the nail beds that ranged in their thickness between 0.8 and 1.0 mm and showed a low velocity flow with a maximum peak systolic velocity of 10.5 cm/s. No arterio-venous shunts were seen in the nail

beds, and the bony margin of the distal phalanx was unremarkable in all cases (Fig. 2).

These sonographic findings support the presence of hyperplasia and/or hypertrophy of the ungual connective tissue and their blood vessels perhaps combined with an inflammatory component. Interestingly, in spite, that the digital arteries were dilated and/or prominent in these cases, the presence of both dilated and numerous arterial vessels was noted mainly within the nail bed tissue and not in the periungual regions or proximal part of the fingers. Thus, we may postulate that perhaps the nail can present a more sensitive response and/or specific receptors to the unbalanced manifestation of the several physicochemical and growth factors involved in the physiopathology of CF which can also be a particular response to the chronic vasodilation of the distal portion of the digital arteries. Additionally, the presence of a prominent ungual hypervascularity may explain the reported presence of splinter haemorrhages in these cases. In conclusion, colour Doppler ultrasound may be a useful imaging technique for understanding the physiopathology of clubbing and exploring '*in vivo*' the nails in this entity.

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References

1 Wenk KS, Higgins KB, Greer KE. Cystic fibrosis presenting with dermatitis. *Arch Dermatol* 2010; **146**: 171–174.
2 Bernstein ML, McCusker MM, Grant-Kels JM. Cutaneous manifestations of cystic fibrosis. *Pediatr Dermatol* 2008; **25**: 150–157.
3 Spicknall KE, Zirwas MJ, English JC III. Clubbing: an update on diagnosis, differential diagnosis, pathophysiology, and clinical relevance. *J Am Acad Dermatol* 2005; **52**: 1020–1028.
4 Rutherford JD. Digital clubbing. *Circulation* 2013; **127**: 1997–1999.
5 Augarten A, Goldman R, Laufer J *et al.* Reversal of digital clubbing after lung transplantation in cystic fibrosis patients: a clue to the pathogenesis of clubbing. *Pediatr Pulmonol* 2002; **34**: 378–380.
6 Holzberg M. The nail in systemic diseases. In Baran R, de Berker D, Holzberg M, Thomas L eds. Baran & Dawber's Diseases of the Nail and Their Management, 4th edn. Wiley-Blackwell, Chichester, UK, 2012: 316–388.
7 Roomans GM, Afzelius BA, Kollberg H, Forslind B. Electrolytes in nails analysed by X-ray microanalysis in electron microscopy. Considerations on a new method for the diagnosis of cystic fibrosis. *Acta Paediatr Scand* 1978; **67**: 89–94.
8 Wortsman X, Wortsman J, Soto R *et al.* Benign tumors and pseudotumors of the nail: a novel application of sonography. *J Ultrasound Med* 2010; **29**: 803–816.
9 Wortsman X. Sonography of the nail. In Wortsman X, Jemec GBE eds. Dermatologic Ultrasound with Clinical and Histologic Correlations, 1st edn. Springer, New York, NY, 2013: 419–476.

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Coexistence of frontal fibrosing alopecia and discoid lupus erythematosus of the scalp in 7 patients: just a coincidence?

Editor

Primary cicatricial alopecias (PCAs) are a rare group of disorders that cause irreversible damage to hair follicles. Classification of PCA is mainly based on the predominant inflammatory cell type.[1] Discoid lupus erythematosus (DLE) and lichen planopilaris (LPP) are forms of PCA with a predominantly lymphocytic infiltrate. Frontal fibrosing alopecia (FFA) is a clinically distinct variant of LPP characterized by a progressive band of alopecia of the frontal hairline.[2] Coexistence of FFA and scalp DLE is supposedly rare, with only one case reported.[3] We describe seven patients presenting such association and discuss the potential common pathogenic background of both conditions.

All patients were women (four Caucasian, three African-descendants; age range 36–68 years; six post-menopausal, one pre-menopausal) complaining both of patchy alopecia and frontal hairline recession from 4 months to 8 years. FFA was the first diagnosis in one patient, DLE in three patients and in three subjects DLE and FFA were diagnosed simultaneously (Figs 1,2c). LPP was also present in one patient and three presented DLE elsewhere in the body (Fig. 2a,b). None had systemic lupus erythematosus (LE). Other features associated with FFA were also

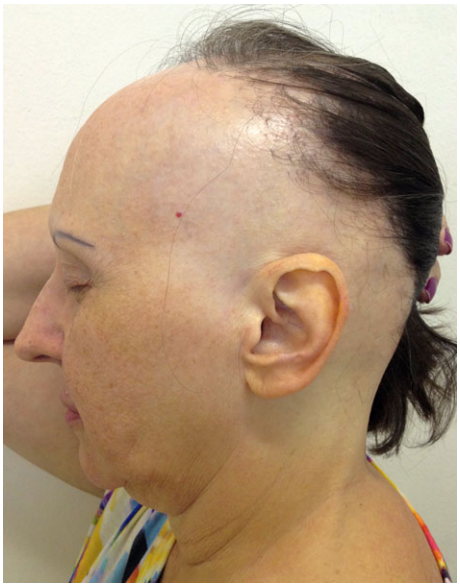

Figure 1 59-year-old patient with advanced frontal fibrosing alopecia with occipital involvement. Facial papules and eyebrow loss are also perceptible.