Letters and comments

Contributors to this section are asked to make their comments brief and to the point. Letters should comply with the instructions to authors at <http://www.rcseng.ac.uk/publications/submissions/authorinstructions.html>. Tables and figures should only be included if absolutely essential and no more than five references should be given. The editor reserves the right to shorten letters and subedit contributions to ensure clarity.

Another epidermal naevus with polyostotic anomalies?

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COMMENT ON

Marsden N, Shokrollahi K, Maw K, Sierakowski A, Bhat FA, Mathur B.

Congenital vascular malformation associated with multiple cranial, vertebral and upper limb skeletal abnormalities. *Ann R Coll Surg Engl* 2010; **92**: online only

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I read with interest the case report by Marsden et al. in which the authors presented an 18-year-old man who was born with a large naevus of the face, neck and shoulder region associated with multiple skeletal abnormalities. The skin lesion was thickened, dark red/brown and was limited by the midline, which the authors referred to later in the report as a port-wine stain (capillary malformation). The authors concluded that the diagnosis is 'congenital vascular malformation associated with multiple skeletal abnormalities'.

I would like to clarify one major point regarding the diagnosis of the skin lesion. The description of the birthmark as thickened, brown and limited by the midline is highly suggestive of a linear epidermal nevus rather than a mere capillary stain. The location and appearance on the published figure also support this alternative diagnosis.

While both capillary malformations and epidermal nevi are congenital stains and may co-exist in the same patient (such as in CLOVES syndrome¹), these two entities are clinically and pathologically distinct with different clinical implications.

Epidermal nevi can be associated with multiple anomalies including skeletal, cerebral, ocular, among others. Examples include several heritable and non-heritable 'epidermal nevus syndromes', including Proteus syndrome, Cowden syndrome, Schimmelpenning syndrome, Becker nevus syndrome and CLOVES syndrome.^{1,2} Various forms of bony anomalies have been reported with epidermal nevi including those similar to fibrous dysplasia.³⁻⁶ None of the disorders mentioned by the authors (Klippel-Trenaunay syndrome, Parkes Weber syndrome and Servelle-Martorell syndrome) typically include an epidermal nevus.

Accordingly, the constellation of cutaneous and skeletal features described by the authors may fall within the spectrum of epidermal nevus syndromes rather than 'a previously undescribed collection of vascular and bony abnormalities' or another illusive 'congenital vascular bone syndrome'.

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