Dear Editors,

We report on a 14-year-old girl who presented with an asymptomatic, rigid, skin-colored and cobblestone-surfaced nodule on her foot. It formed at the age of four years and had not grown further until a few months before presentation. The nodule was located on the right sole of her foot and was accompanied by several coalescing papules on the medial nailfold of her great toe and the adjacent interdigital space (Figure 1a, b). The family history was unremarkable.

Two biopsies showed an unremarkable epidermis and underlying coarse, homogeneous collagen bundles with strong focal mucin deposits (Figure 2). Fibroblasts and elastic fibers were diminished. Staining for CD68 showed scattered histiocytes. We performed immunohistochemical staining to rule out common soft tissue tumors. Actin and desmin stains were negative; this excluded the presence of myofibroblasts that are commonly found in cell-rich dermatofibromas. Desmin staining was also negative and the absence of inclusion bodies excluded infantile digital fibromatosis (a benign myofibrocytic tumor that appears on the fingers and toes in early childhood) [1]. Staining for CD34, typically positive in dermatofibrosarcoma protuberas, was also negative. The patient was diagnosed with a plantar collagenoma – a hamartoma of the connective tissue nevi family (Figure 3a) [2, 3].

Collagenomas typically show an accumulation of dense, coarse, hyalinized collagen and a normal or diminished quantity of elastic fibers in the dermis. The epidermis does not show characteristic changes [2]. The cytomorphology of sparse dermal spindle cells is usually unremarkable, although collagenomas occasionally show giant cells (giant cell collagenomas). The considerable mucin accumulation in our case is not a typical characteristic of collagenomas but has been described in other cases of isolated collagenoma [4]. We therefore...
interpreted it as a morphological variant. Malignant soft tissue tumors that are relatively prevalent on the extremities, such as angiomatoid fibrous histiocytoma and malignant peripheral nerve sheath tumor, were excluded histologically [5, 6].

Collagenomas may be isolated or accompany congenital diseases (Figure 3b). The inheritance pattern of these congenital diseases is either autosomal dominant (familial cutaneous collagenoma syndrome, MEN-1, Buschke-Ollendorf syndrome and tuberous sclerosis) or sporadic (Proteus syndrome). As collagenomas can develop early in the course of a congenital disease, they have an important signaling function for the treating dermatologist.

Plantar collagenomas are a common finding in Proteus syndrome (PS). Plantar collagenomas with a cerebriform

Figure 2  Histological findings. Hematoxylin-eosin staining showing an unremarkable epidermis with underlying coarse, homogeneous collagen bundles and mucin deposits. Fibroblasts are slightly diminished (a, b). Staining of elastic tissue shows thick collagen bundles and diminished elastic fibers (c).

Figure 3  Classification of connective tissue nevi. Classification of connective tissue nevi (graphic adapted from [3]) (a). Classification of congenital and acquired collagenomas (graphic adapted from Uitto et al. [2] and McClung et al. [11]) (b).

Abbrev.: FCC SD, familial cutaneous collagenoma syndrome; MEN1, multiple endocrine neoplasia type 1; SD, syndrome
surface are nearly pathognomonic for PS. The mutation responsible for the syndrome is strictly sporadic and causes a somatic mosaicism. The mutation leads to activation of the AKT1 oncogene, which is responsible for cell proliferation and apoptosis. It leads to disproportionate and progressive overgrowth of the affected tissue, including skin, bones and internal organs [7, 8]. Depending on the affected organ, symptoms of PS are highly variable; it was therefore named after the Greek shape-shifting god Proteus.

Acquired collagenomas are not associated with any of the above-mentioned syndromes and may present as eruptive collagenoma (multiple nodules) or as an isolated collagenoma (a single nodule) that appears in early childhood. All of the described types of collagenoma show similar histological findings.

In our patient, clinical examination and sonography of the abdomen and ovaries did not show any signs of PS and the patient was therefore diagnosed with isolated plantar cerebriform collagenoma, without associated clinical abnormalities.

The etiology of acquired collagenoma is not yet fully understood. Uitto et al. showed that excessive accumulation of collagen type 1 in patients with isolated collagenoma is partly caused by decreased collagenase production and faster fibroblast multiplication in the lesion than non-affected skin of the same patient [9, 10].

In mild cases, treatment is not necessary [11]. If required, treatment options include excision [12] and repeated intralesional steroid injection [13]. Our patient was treated with a partial excision to avoid a free flap transplant. The major part of the tumor was excised and subsequently treated with a collagen-glycosaminoglycan bilayer wound dressing combined with negative pressure wound therapy. After 23 days, the wound dressing was removed and a split skin transplant from the right sole was performed on the left sole. On the toe and the interdigital area, a full skin transplant from the left groin was performed. At the six-month follow-up appointment, wound healing was satisfactory and further excision was not necessary.

In conclusion, we report a detailed case of a young patient with an isolated plantar collagenoma that was successfully treated with surgery. Clinicians should be familiar with the associated syndromes in order to exclude a collagenoma.

Conflict of interest
None.

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