Onychomatricoma with Concomitant Subungual Glomus Tumor

Penelope Kallis · Mariya Miteva · Tejas Patel · Martin Zaiac · Antonella Tosti

Department of Dermatology and Cutaneous Surgery, University of Miami Miller School of Medicine, Miami, Fla., USA

Established Facts

- Onychomatricoma is a benign nail tumor.
- Glomus tumor is a painful benign nail tumor.

Novel Insights

- We describe the association of these two tumors on the same nail.

Key Words

Glomus tumor · Nail pathology · Nail tumor · Onychomatricoma

Abstract

Onychomatricoma and glomus tumor are two rare subungual neoplasms with distinct clinical and histopathological features. We report a case of onychomatricoma associated with a glomus tumor in the subungual region of the same finger in a 45-year-old woman. Histopathological examination revealed characteristic findings of both onychomatricoma and glomus tumor. To the best of our knowledge, these two subungual tumors have never before been described occurring concomitantly.

© 2015 S. Karger AG, Basel

Introduction

Onychomatricoma is a benign fibroepithelial tumor of the nail matrix characterized histologically by its projections into the attached nail plate [1] and clinically by a thickened and yellow discolored nail plate [2]. Subungual glomus tumors are painful vascular neoplasms arising from glomus bodies [3, 4] that result in red or blue discoloration of the nail plate [5]. This report documents the clinical and histopathological features of a novel case of a patient with both onychomatricoma and subungual glomus tumor of the left fourth fingernail.

Case Report

A 45-year-old woman presented with a 10-year history of a thickened and discolored left fourth fingernail. Her only symptom involved pain at compression. She denied a history of trauma to the area. Examination of the nail revealed a purple-blue area of discoloration proximally as well as a mild thickening and yellow

Fig. 1. a A purple discolored proximal nail plate with a thickened overcurved distal margin containing splinter hemorrhages and cavitations (color refers to the online version only). **b** Nail dermoscopy reveals proximal purple discoloration, longitudinal white

lines and distal splinter hemorrhages. **c** Frontal view of the thickened nail plate with honeycomb cavities at the distal margin. **d** Surgical avulsion showed a tumor appearing from the nail matrix.

discoloration of the distal nail plate (fig. 1a). Dermoscopy showed proximal purple discoloration as well as white lines and splinter hemorrhages in the distal nail (fig. 1b). The frontal view revealed holes in the distal margin of the nail plate (fig. 1c).

The nail plate was avulsed, allowing a subungual neoplasm to be exposed and excised (fig. 1d). Both the nail plate and tumor specimens were sent to pathology for microscopic evaluation. Longitudinal sections of the specimens were stained with hematoxylin and eosin, periodic acid-Schiff and for smooth muscle actin prior to examination.

On the basis of the clinical and pathological findings, a diagnosis of onychomatricoma and subungual glomus tumor was established (fig. 2). Histopathological analysis of the neoplasm revealed

a circumscribed nonencapsulated dermal nodule composed of convoluted dilated vascular channels surrounded by layers of cuboidal basaloid cells of the same size (fig. 3). The specimen also stained strongly positive for smooth muscle actin, and the nail matrix was present but with a thicker keratogenous zone. Digitate epithelial proliferations were noted extending from the nail matrix and perforating the nail plate along its longitudinal axis (fig. 4). A transverse section of the dystrophic nail plate revealed that the projections formed cavities filled with serous material. Layers of the matrix cells invading the nail plate and cavities with fluid were also observed on the nail clipping from the distal nail plate obtained at the time of excision. The periodic acid-Schiff stain for fungal organisms was negative in both specimens.

Fig. 2. Longitudinal section revealing concomitant onychomatricoma (left) and glomus tumor (right) (hematoxylin and eosin stain; original magnification $\times 2$).

Fig. 3. Layers of similarly sized cuboidal basaloid cells surrounding convoluted dilated vascular channels (hematoxylin and eosin stain; original magnification $\times 4$).

Fig. 4. Longitudinal section illustrating digitate epithelial projections extending from the nail matrix and perforating the proximal nail plate (hematoxylin and eosin stain).

Discussion

Onychomatricoma is a benign tumor that originates from the nail matrix [2]. It most frequently affects the middle-aged population, with no difference in prevalence amongst sexes [6]. Hallmark features include a thickened yellowed nail plate, overcurvature [2] and multiple channel-like cavities in the distal edge [1].

A diagnosis of onychomatricoma can be made based on its clearly defined histological characteristics. Two zones demarcate the pedunculated fibroepithelial tumor. The base of the tumor is located beneath the proximal nail fold in the proximal zone, and it is exposed following nail avulsion [7, 8]. The proximal zone is lined with papillomatous epithelial invaginations and characteristic thick V-shaped keratogenous zones mimicking the normal nail matrix and distally resulting in nail thickening [8]. The epithelium overlays a biphasic stroma composed of a fibroblast-rich superficial layer and a deep layer that is markedly hypocellular and collagenous [2, 8]. Unlike other nail tumors that indirectly impact the nail plate, onychomatricoma is the only nail tumor of which the lesion itself produces nail plate alterations [9]. The distal zone is associated with the lunula and can be described as multiple 'glove-finger' projections that perforate the attached nail plate [8]. Because of its unique involvement with the nail plate, a diagnosis of onychomatricoma can also be done by histological examination of nail clippings. A nail specimen reveals serous fluid-filled cavities lined with a thin layer of matrix epithelium, representing the digitations of the distal zone [1].

Glomus tumor is a rare benign neoplasm of the glomus body, a neuromyoarterial structure that functions in thermoregulation [3]. Glomus bodies reside in the dermis in multiple locations in the body, but they are particularly concentrated in the subungual regions of the fingertips [3]. Thus, glomus tumors occur most commonly in the

subungual location, and this location has been especially associated with middle-aged women [4]. The classic triad of symptoms includes intense, paroxysmal pain, pinpoint tenderness and cold sensitivity [4]. Glomus tumor frequently presents as a reddish or bluish macule visible underneath the nail plate [4, 5].

Pathologically, glomus tumors are well circumscribed and usually no larger than 1 cm [10]. They have been described as hamartomas with the proliferation of glomus cells, smooth muscle cells and vascular components of the normal glomus body [11]. Depending on the relative concentrations of these three entities, glomus tumors can be classified into solid glomus tumors, glomangiomas or glomangiomyomas [3]. Uniformly round or ovoid glomus cells localize around blood vessels [3, 5]. Their cytoplasm is slightly eosinophilic, and their borders are clear but may be enhanced with a reticulin or periodic acid-

Schiff stain [3]. On immunohistochemistry, glomus tumors stain positive for antibodies to vimentin, smooth muscle actin and CD34 [10].

The differential diagnosis for onychomatricoma includes fungal infections of the nail, benign tumors such as fibrokeratoma, periungual fibroma and verruca vulgaris, and malignant tumors such as Bowen's disease [12]. Painful tumors of the digits should be included in the differential diagnosis for glomus tumors, such as neuroma, leiomyoma, ganglion or exostosis as well as other painful conditions such as gouty arthritis [5]. To date, no case of onychomatricoma and glomus tumor comanifesting in the same patient has been described in the literature. Onychomatricoma has been found with coexisting onychomycosis [13], while glomus tumors have not been reported with any other nail pathology until now.

References

- 1 Miteva M, de Farias DC, Zaiac M, Romanelli P, Tosti A: Nail clipping diagnosis of onychomatricoma. *Arch Dermatol* 2011;147:1117-1118.
- 2 Baran R, Kint A: Onychomatrixoma. Filamentous tufted tumour in the matrix of a funnel-shaped nail: a new entity (report of three cases). *Br J Dermatol* 1992;126:510-515.
- 3 Gombos Z, Zhang PJ: Glomus tumor. *Arch Pathol Lab Med* 2008;132:1448-1452.
- 4 Van Geertruyden J, Lorea P, Goldschmidt D, de Fontaine S, Schiund F, Kinnen L, Ledoux P, Moermans JP: Glomus tumours of the hand. A retrospective study of 51 cases. *J Hand Surg Br* 1996;21:257-260.
- 5 Samaniego E, Crespo A, Sanz A: Key diagnostic features and treatment of subungual glomus tumor (in Spanish). *Actas Dermosifiliogr* 2009;100:875-882.
- 6 Rashid RM, Swam J: Onychomatricoma: benign sporadic nail lesion or much more? *Dermatol Online J* 2006;12:4.
- 7 Perrin C, Baran R, Pisani A, Ortonne JP, Michiels JF: The onychomatricoma: additional histologic criteria and immunohistochemical study. *Am J Dermatopathol* 2002;24:199-203.
- 8 Perrin C, Goettmann S, Baran R: Onychomatricoma: clinical and histopathologic findings in 12 cases. *J Am Acad Dermatol* 1998;39:560-564.
- 9 Haneke E, Fränken J: Onychomatricoma. *Dermatol Surg* 1995;21:984-987.
- 10 Kale S, Rao V, Bentz M: Glomus tumor of the index finger. *J Craniofac Surg* 2006;17:801-804.
- 11 Carroll RE, Berman AT: Glomus tumors of the hand: review of the literature and report on twenty-eight cases. *J Bone Joint Surg Am* 1972;54:691-703.
- 12 Baran R, Richert B: Common nail tumors. *Dermatol Clin* 2006;24:297-311.
- 13 Fayol J, Baran R, Perrin C, Labrousse F: Onychomatricoma with misleading features. *Acta Derm Venereol* 2000;80:370-372.