Clinical Case Report

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Nail bed solitary neurofibroma

A case report and literature review

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Abstract

Rationale: Nail bed solitary neurofibroma is an extremely rare tumor, with only 9 cases recorded in the literature so far.

Patient concern: We present the case of a 42-year-old female patient, with a history of a slowly growing tumor in the nail bed of the left index and no clinical features of type I neurofibromatosis.

Diagnosis: Nail bed solitary neurofibroma.

Intervention: The tumor was surgically removed and the pathology examination established the diagnosis of neurofibroma.

Outcomes: The postoperative outcome was good, with no recurrence 12 months after surgery.

Lessons: We present the rarity of this type of tumor localized in the nail bed, taking into consideration the only 9 cases recorded in the literature. Nail bed solitary neurofibroma should be also included in the differential diagnosis of a nail bed tumor.

Abbreviation: MRI = magnetic resonance imaging.

Keywords: nail bed neurofibroma, pathology examination, surgery

1. Introduction

Neurofibromas are benign nerve-sheath tumors arising from Schwann cells and account for 5% of all benign soft tissue neoplasms.[1] They are usually solitary tumors but in about 10% of cases are found in association with neurofibromatosis type I. Neurofibromas can arise anywhere along a nerve fiber and usually involve nerves in or near the skin. They rarely develop in the hand and may affect the digits. However, nail bed solitary neurofibroma is an exceptionally rare event with no more than 9 cases described in the literature.[2–10] We herein report a rare case of nail bed solitary neurofibroma in the left second finger. A review of the cases published in the literature is also presented.

2. Case report

We present the case of a 42-year-old female patient with a tumor located in the radial side of the nail bed of the left-hand second finger. The white–pink colored solid tumor was circumscribed, had a nodular shape, with a diameter of approximately 1 cm, painless spontaneously or on palpation, but with significant onychodystrophy. Thinning and raising on the radial side of the nail plate was noticed (Fig. 1). The age of the tumor at the time of diagnosis was approximated by the patient at 2 years, with a slow growth and no history of trauma. The patient did not report any symptom, the only reason for seeking medical advice being nail esthetics, significantly affected. No clinical features of neurofibromatosis type I were found. The patient completed and signed the informed consent. Radiological examination revealed bone distortion by compression of the distal phalanx of the left second finger, without cortical damage or bone invasion (Fig. 2). Ultrasound examination described a hypoechogenic nodular tumor of 11 mm in size with a polycyclic profile and low Doppler signal. Surgery was performed under local anesthesia. Intraoperatively, after raising the nail plate, a well-circumscribed

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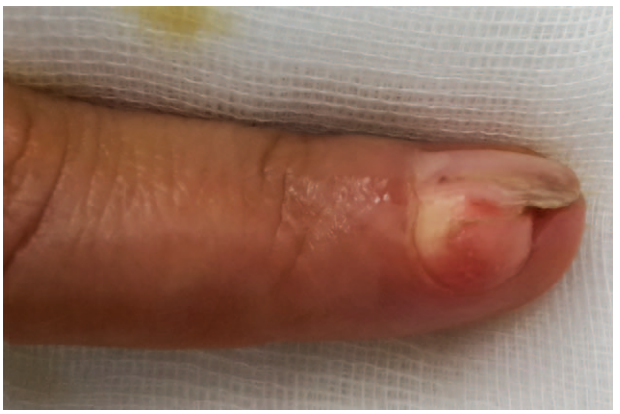

Figure 1. Nail bed solitary neurofibroma - nodular 1 cm white–pink circumscribed solid tumor with important onychodystrophy in the radial side of the left 2nd finger nail bed.

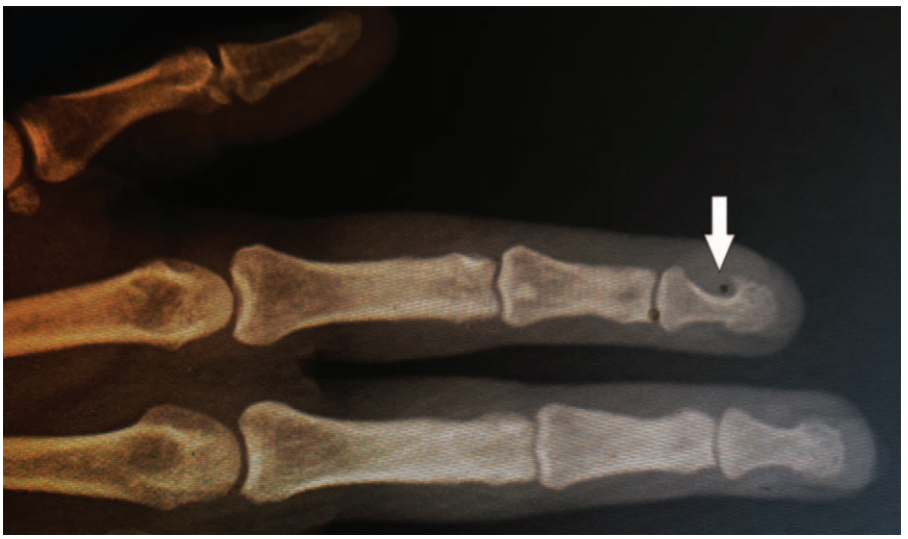

Figure 2. Bone distortion without cortical damage or bone invasion.

white, smooth, and relatively soft tumor, expanding toward the pulp of it was seen. The excision was simple and left a relatively large defect. The specimen size was 11/15 mm. Direct suture with nail bed repositioning was performed.

The histopathological exam revealed the presence of a relatively well-defined tumor proliferation, consisting of bundles of fusiform cells with elongated nuclei, without cytonuclear pleomorphism and mitotic activity. There were also myxoid areas, thick collagen bands and reduced inflammatory infiltrate (Fig. 3). All these microscopical aspects were suggestive of neurofibroma. Immunohistochemical staining for S100 protein was positive (Fig. 4). The follow-up revealed no local recurrence 12 months after surgery.

3. Discussion

Neurofibroma has been mainly reported in the peripheral nerves of the lower and upper limbs and may be solitary or part of a more general neurofibromatosis. However, nail bed solitary neurofibroma is a rare event. Runne et al and Shelly et al[11,12] are credited with the first two cases of a subungual neurofibroma reported in the literature, but without any data regarding the presence or absence of Recklinghausen disease. Until now, only 9 case reports of nail bed solitary neurofibroma have been documented (Table 1).

Nail bed solitary neurofibroma occurs either in the upper limb (7 out of 10 cases including ours) or lower limb (3 patients). They tend to be small tumors (range 0.5 cm to 1.8 cm) with nonspecific clinical features.[3,8] These tumors are slow growing and rarely painful. No complaint of pain was recorded in all cases but 1, our case included. The longest history was 6 years (average 2.4 years) for a left thumb tumor in a 60-year-old woman.[5] Middle-aged women with 8 out of all 10 cases seem to be more affected by this type of tumor (Table 1). Nail bed solitary neurofibroma may cause nail plate deformity, onychodystrophy, or hyperkeratosis. Thickening and elevation of the nail plate were described in 3 cases while onychodystrophy was mentioned in the other 3 cases (Table 1). There was 1 case with no nail plate deformity.[8] Due to these features nail bed solitary neurofibroma is difficult to diagnosis. Other tumors such as glomus tumor, Koenen tumor, fibrokeratoma, or squamous cell carcinoma may clinically mimic

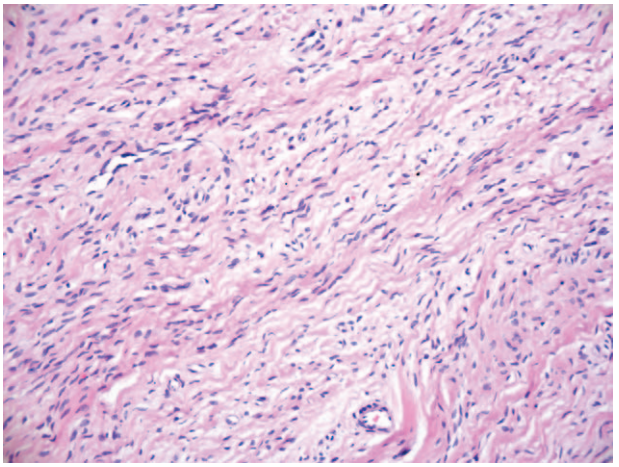

Figure 3. Optical microscopy of nail bed neurofibroma showing slightly wavy, elongated cells with discrete angled nuclei and interstitial collagen fibers (hematoxylin-eosin ×20).

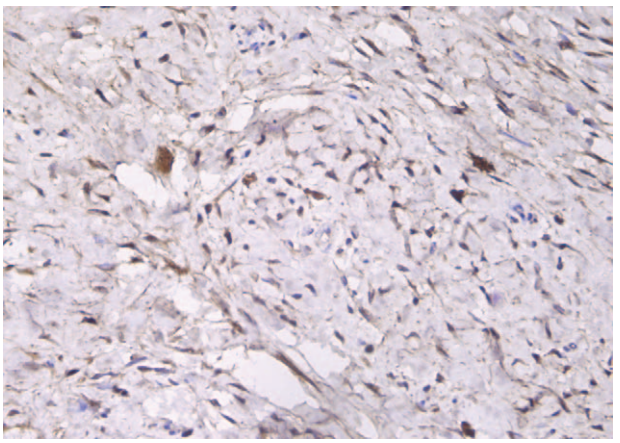

Figure 4. Immunochemistry examination: S100 protein staining positive in tumor cell nuclei.

Table 1

Cases of nail bed solitary neurofibroma published in the literature to date.

	Author year	Location	Age years	Sex	History	Symptoms	Clinical findings	Radiology	Size (cm)	Recurrence
1	Niizuma[2] 1991	Right third finger	13y	F	2y	No pain	Deformed overall nail into a flattened dome shape	Mild compression atrophy	_	No at 17 m
2	Bhushan[3] 1999	Right third finger	27y	F	2y	No pain	Thickening and elevation of the nail; Subungual hyperkeratosis and diffuse, nonfluctuant swelling beneath the proximal nailfold	Soft tissue swelling with no focal bone or joint changes	0.5 × 0.5	–
3	Dangoisse[4] 2000	Right hallux	43y	F	4y	Slight pain on direct pressure	Onychodystrophy; subungual mass; blue-red discoloration of the proximal nail plate	Bone print without bony invasion	1.0 × 1.5	no
4	Baran[5] 2001	Left thumb	60y	F	6y	No pain	Nail deformity with marked transverse overcurvature	Normal	–	–
5	Sugiur [6] 2004	Left third toe	50y	F	–	–	Tumor	No bone defects	–	No at 6 m
6	Roldan-Marin[7] 2006	Right thumb	50y	F	18m	Pain	Thickening and elevation of the nail plate; paronychia and onychodystrophy	–	–	No at 9 m
7	Stolarczuk[8] 2011	Right fourth toe	51y	M	8m	No pain	Smooth, rounded, regular, normochromic tumor	No bone abnormality Hypoechoic nodule with no flow at Doppler	1.6 × 1.8	No at 6 m
8	Bom Mie Seo[9] 2011	Right thumb	44y	M	3y	No pain	Onychodystrophy and nail plate elevation; lunula distorsion	–	1.0 × 0.8	No at 14 m
9	Huajun[10] 2012	Right first finger	32y	F	2y	No pain	Thickening and elevation of the nail plate	No bone defects Hyperintense T2-weighted mass occupying subungual area	1.0 × 1.0	No at 10 m

F = female, M = male, m = months, y = years.

neurofibroma and must be included in the differential diagnosis.[12,13] Some authors consider that these tumors are sometimes misdiagnosed as onychomycosis or even trauma leading to underdiagnosis of these lesions.

Imaging tools are not very helpful in making the diagnosis, in 5 out of 9 cases radiology being normal. In 2 formerly reported patients and also in our patient bone print without bony invasion was noted (Table 1). An ultrasound exam cannot reliably distinguish schwanomma from neurofibroma, the sonographic features being almost identical – moderately echogenic solid elongated mass and generally with no Doppler sign or contrast enhancement.[14] In 1 case ultrasound exam showed a hypoechoic mass without Doppler flow, while in our case a hyperechoic mass with low-intensity Doppler flow was found.[8] Magnetic resonance imaging (MRI) appearance of neurofibromas is nonspecific, similar to that of schwannomas. They generally exhibit hypo- to isointense signal to muscle on T1-weighted images and hyperintense signal on T2-weighted sequences, with a variable enhancement pattern and foci of heterogenity.[15] Due to the small tumor size, peripheral location, and nonspecific imaging features of nail bed solitary neurofibroma MRI was rarely used for preoperative diagnosis. In only one case MRI was used and the exam characterized neurofibroma as a hyperintense mass on T2-weighted image.[10] Due to the nonspecific clinical presentation and radiological features, surgery is essential in establishing the diagnosis. At surgery nail bed solitary neurofibroma is a solid, relatively well-circumscribed, non-encapsulated, soft, or firm tumor. In all described cases, after nail plate removal (partial or complete) the tumor is easily removed, which was also the case with our patient (Table 1). In none of the reported cases, plastics for the reconstruction of the nail bed or marginal edges were used. Patient satisfaction, in the presented case, was maximum.

Final diagnosis rests on the pathology exam. In all cases, histopathology examination revealed well-circumscribed non-encapsulated tumor consisting of interlacing neural filaments bundles of loosely spindle cells with elongated nuclei and pale cytoplasm in a myxoid stroma. However, pathology examination cannot always differentiate neurofibroma from other tumors in this location, like schwannoma or solitary fibrous tumors. Immunochemistry may help to show staining for S100 protein. Staining for S100 is also present in schwannoma but is more intense and uniform than in neurofibroma, while in fibrous tumors staining for S100 protein is negative and for CD 34 strongly reactive.[10] In 1991, Niizumaet et al[2] used electron microscopy to analyze their case of nail bed solitary neurofibroma, and found that Schwann cells and fibroblasts accounted for several percents of the total cells, and parenchymal cells, equivalent to perineurial cells, accounted for more than 90% of the cells, and staining for protein S100 was positive in more than 90% of the parenchymal cells confirming the presence of Schwann cells.

No recurrences were recorded after surgical removal in any published case of nail bed solitary neurofibroma with a maximum follow-up of 17 months.[2]

Despite the rarity of this type of tumor, nail bed solitary neurofibroma with its nonspecific physical and imaging features must be included in differential diagnosis of nail bed tumors, a high index of suspicion contributing to a better diagnosis.

Complete surgical excision should be considered in all cases, as a diagnostic and curative tool.

Author contributions

Conceptualization: Mihaela Pertea and Sorinel Lunca.
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Investigation: Mihaela Pertea, Cristina Terinte, Vladimir Poroch, and Natalia Velenciuc.
Writing – original draft: Mihaela Pertea, Oxana-Madalina Grosu, and Sorinel Lunca.
Writing – review & editing: Mihaela Pertea and Sorinel Lunca.

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