4 Advances in MRI of Glomus Tumors of the Fingertips

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Because of their highly characteristic clinical symptoms, glomus tumors are probably the best-known nails tumors. Glomus tumor is a benign and vascular hamartoma that originates from the neuromyoarterial cells of the glomus bodies (Masson's glomus) in the reticular dermis, most often at the nail bed. Many recent studies have provided refinements in imaging features and imaging strategies.

Clinical Exam

Glomus tumors occur more frequently in women between 30 and 50 years old. Clinical diagnosis is based on the classic triad of cold hypersensitivity, pinpoint tenderness (Love's test), and paroxysmal pain. The pain can radiate up to the elbow or even the shoulder. A tourniquet at the base of the finger or a blood pressure cuff inflated to 300 mm Hg is able to alleviate the pain. Cold hypersensitivity is not the rule and noted between 31% and 42% of the patients [1, 2]. Some rare painless cases are reported [3]. A bluish or violaceous spot of the lunula or the nail bed and a reddish line extending distally may be present in 43% of cases (Fig. 4.1) [4]. The nail may be slightly elevated over the lesion or even split. The classic subungual location seems lower (55%) than that previously published (75%–90%) [2, 5]. Rare cases of glomus tumor originating directly from a digital nerve are reported [6, 7]. Infection on a ruptured tumor is exceptional [8]. Physical examination and medical history may be informative enough for a correct diagnosis. However, the mean delay for the diagnosis needs usually several years. Histology consists of a convoluted

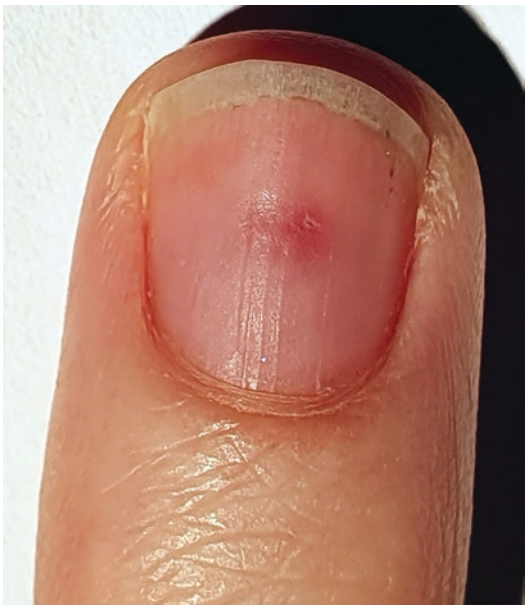

Fig. 4.1 Reddish spot at the midline of the nail bed of the fourth finger of a 38-year-old female

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R. L. Baran (ed.), *Advances in Nail Disease and Management*, Updates in Clinical Dermatology,
https://doi.org/10.1007/978-3-030-59997-3_4

arteriovenous anastomosis with a relative paucity of a vascular lumen surrounded by a thick layer of modified smooth muscle cells and nerve elements. Surgical excision is the most effective treatment.

MRI

The role of imaging during the management of a glomus tumor is poorly helpful for visible lesions with characteristics symptoms [2]. Radiographs present a low sensitivity and specificity with bone erosion of the dorsal cortex of the distal phalanx in 36% of cases [9]. Ultrasound can detect tumors as small as 2 mm but remains strongly dependent on the expertise of the operator (but as well as for MRI).

These last year's numerous studies assessed not only the accuracy but also the insufficiencies of MRI to diagnose subungual glomus tumors. MRI is useful when the location of the tumor is in doubt or in the case of suspected multifocal lesions [10].

MRI is more accurate than plain films to depict associated bone erosion of the distal phalanx. Usually, the lesion is well defined with sometimes a pseudocapsule. The signal is homogeneous iso or slightly intense on T1 WI, highly intense on T2 WI, and highly enhanced after intravenous injection of gadolinium (Fig. 4.2). However, in some cases, the

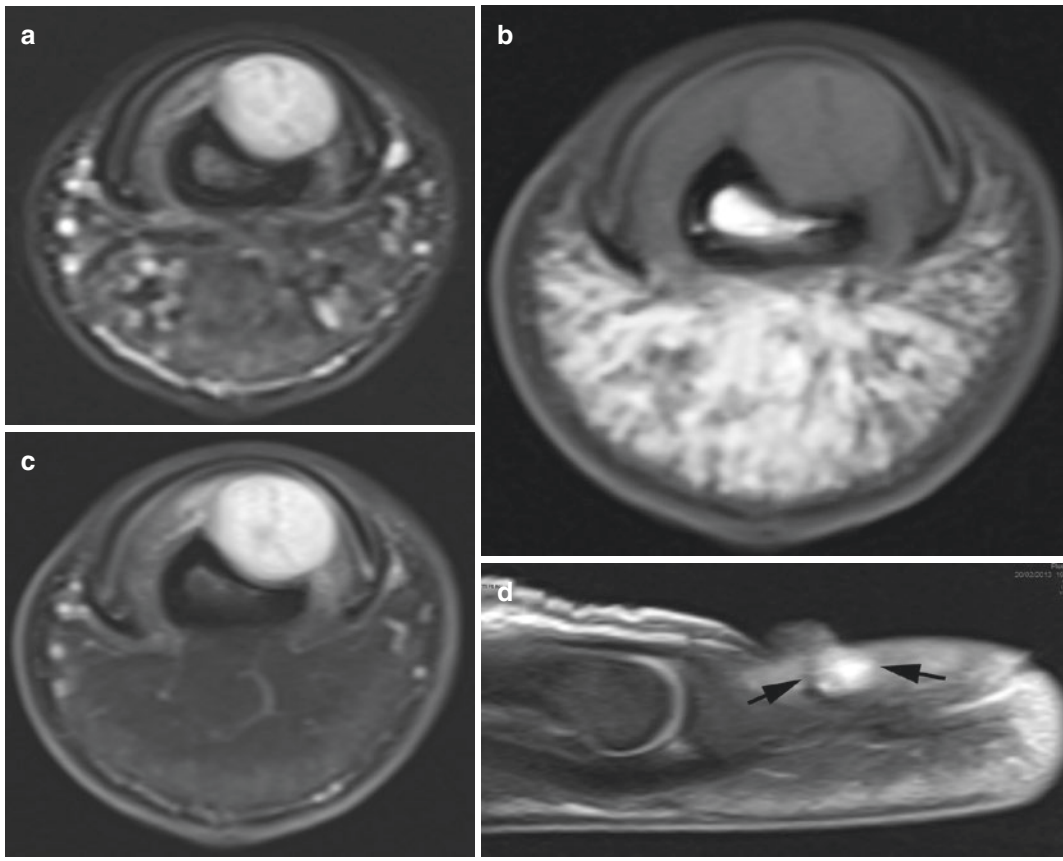

Fig. 4.2 Typical glomus tumor of the left third finger of a 32-year-old female. (**a**) Axial T2 fat-sat WI, (**b**) axial T1 WI, (**c**) axial, and (**d**) sagittal T1 fat-sat WI after intravenous injection of gadolinium. Typical patterns with well-defined limits and ahigh contrast between the glomus tumor and the dermis of the nail bed: high signal on T2 WI, slightly high signal on T1 WI, and strong enhancement. Deep bone erosion of the dorsal cortex of the distal phalanx. The tumor (arrows) is distal to the matrix on the sagittal slice

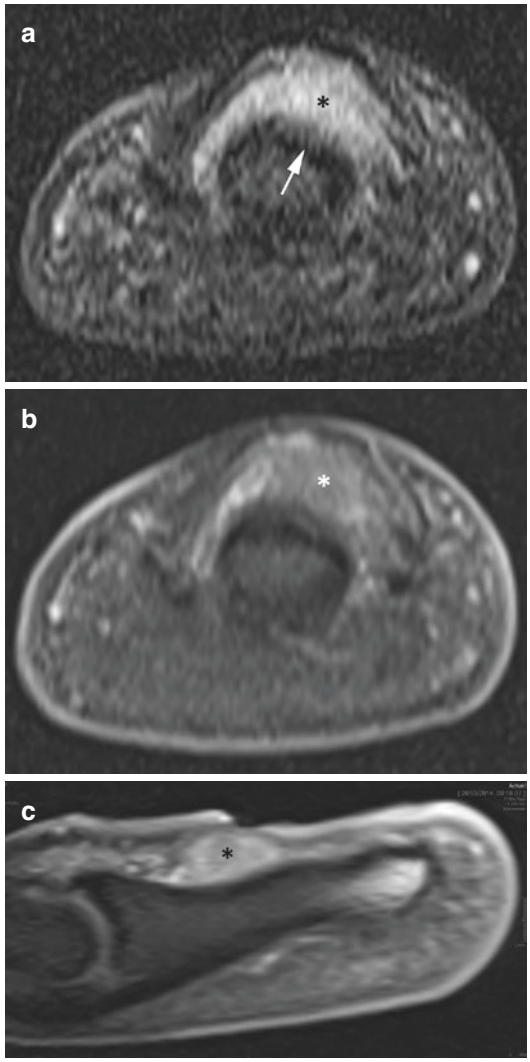

Fig. 4.3 Atypical glomus tumor of the right fifth finger of a 42-year-old female with slight enhancement. (**a**) Axial STIR and axial (**b**) and sagittal (**c**) T1 WI after intravenous injection of gadolinium. The tumor is difficult to highlight with a signal close to the signal of the surrounding submatrical dermis. Indirect signs are the dorsal shift of the matrix and the superficial bone erosion of the dorsal cortex of the phalanx (arrow)

degree of enhancement on T1 post-contrast imaging may be mild to moderate, remaining isointense to the surrounding enhanced nail bed (Fig. 4.3). The PPV of MRI is high 97% in the diagnosis of glomus tumors as small as 2 mm among 42 patients [11] and 100% among 21 patients [12]. However, MRI presents a low specificity and low NPV: a negative image does not rule out a glomus tumor [1, 12]. Among all patients, 10% have a nondiagnostic MRI [11]. Technical insufficiencies of MRI (misfit coil or protocol, too low spatial resolution) may explain these negative exams. This may be also due to the lack of detection of the smallest tumors on MRI, and surgical exploration may be performed despite a negative MRI. Over time, tiny glomus tumors may become visible and detectable with a delayed MRI (Figs. 4.4 and 4.5). In solid glomus tumors with relatively low vascular lumens, the signal may be similar to the surrounding tissue and can be more difficult to detect (Fig. 4.3). Some lateral location may be difficult to detect on MRI (Figs. 4.5 and 4.6). Submatrical dermis usually presents a high vascularization and may occult a glomus tumor in his area (Fig. 4.7). Performances of MRI may be improved with enhancement curve and/or MR angiography showing an early and partial enhancement at the arterial phase with a progressive and complete enhancement on more delayed acquisitions (Figs. 4.8 and 4.9). A delayed washout of the glomus tumor is possible (Fig. 4.10). On the other hand, potential lesions that exhibit focal high signal only on T2 sequences but without correlative findings on other sequences are more likely to be false positives (Fig. 4.11) [13].

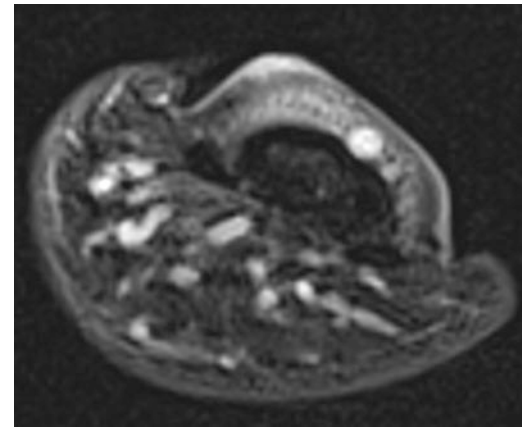

Fig. 4.4 Tiny glomus tumor of the right thumb of a 57-year-old female. Axial T2 fat-sat WI with a 1.3-mm large tumor in the deep dermis of the nail bed lying of the dorsal cortex of the phalanx. A high spatial resolution is necessary to depict such a small lesion

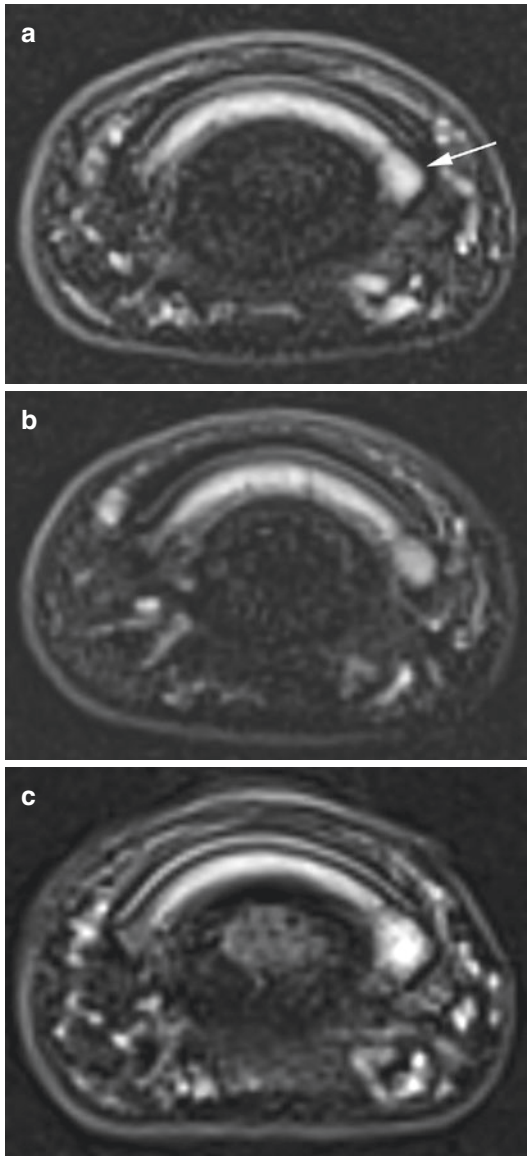

Fig. 4.5 Follow-up of a tiny glomus tumor of the left finger of a 26-year-old male. Axial T2 fat-sat WI in (**a**) 2014, (**b**) 2015, and (**c**) 2019. The tumor is located in the lateral part of the nail bed close to the rima ungalum. The initial size of the tumor was 1.5 mm and reached 2.5 mm 5 years later

However, in more complicated scenarios like suspicion of recurrence after surgery, the predictive power of the clinical exam declines, and the usefulness of MRI increases. In these cases, MRI provides a greater sensitivity than the clinical exam but need a perfect technical protocol with MR angiography. An associated Raynaud's syndrome or vasculitis may disturb the quality of MR angiography (Fig. 4.12). A warm bath of the hands a few minutes before the exam can improve the dynamic injection of chelate of gadolinium.

Glomangioma

Glomus tumors are commonly solitary subungual lesions of the nail bed. Histopathologically, based on the predominant tissue type present, glomus tumors are classified as solid glomus tumors, glomangiomas, or glomangiomyomas. Vascular forms of glomus tumors or glomangiomas (10%–25% of glomus tumors) have a different clinical presentation and are usually multifocal, bluish, painless, and extradigital [14]. Roughly 10% of glomus tumors are multiple, which may be the cause for presumed recurrence (Fig. 4.13). Glomangiomas in subungual location are rare and do not often show the classic triad of symptoms associated with glomus tumors [15]. They can extend to the periungual skin. They can often be misdiagnosed as vascular malformations, resulting in delayed diagnosis and inappropriate treatment. Histological examination is necessary to rule out a clinically indistinguishable benign or malignant melanocytic tumor (blue nevi or melanomas), a venous malformation, or a blue rubber bleb angiomatosis [14]. Glomangiomas are distinguished from solid glomus tumors by the predominant vascular component and less defined limits.

Glomus Tumors and Neurofibromatosis Type 1

Glomus tumors have recently been reported in individuals with neurofibromatosis type 1 (NF1) [4]. They frequently appear as bluish

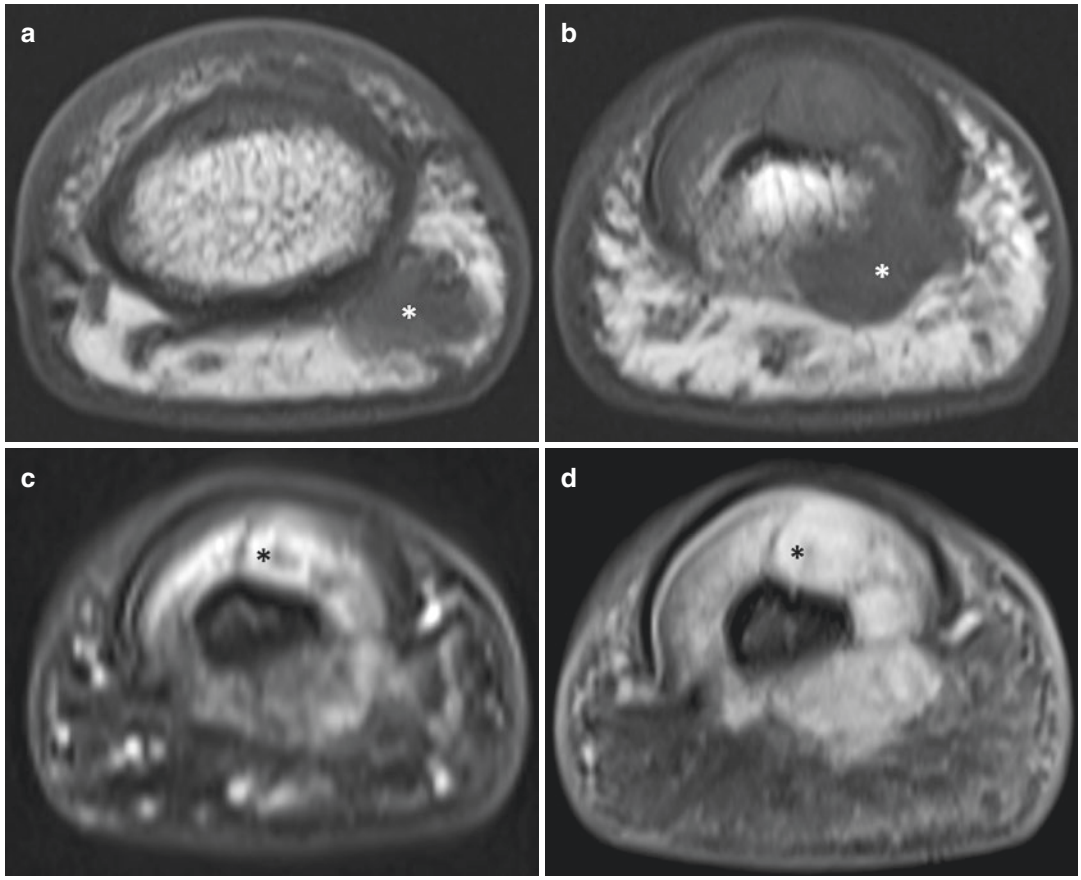

Fig. 4.6 Ill-defined glomus tumor of the midlateral nail bed with extension to the pulp of the right thumb of a 44-year-old female. (**a**, **b**) Axial T1 WI: the tumor extension to the pulp (*) is highlighted by the surrounding fatty tissue. (**c**) Axial T2 WI and (**d**) T1 fat-sat after intravenous injection of gadolinium: the tumor invasion extends to the midline of the nail bed (*)

subcutaneous nodules on the trunk and limbs and can be multifocal [13]. Multifocal tumors (16.7%) and tumor recurrence (33.3%) are more common in association with NF1 than in sporadic cases. There is no correlation between café-au-lait macule burden and the number of neurofibromas with the development of glomus tumors. NF1-associated glomus tumors exhibit no neurofibromin immunoreactivity, whereas their sporadic counterparts retained neurofibromin expression. Detection of glomus tumors and particularly multiple glomus tumors should raise suspicion for a concurrent diagnosis of NF1 (Fig. 4.14).

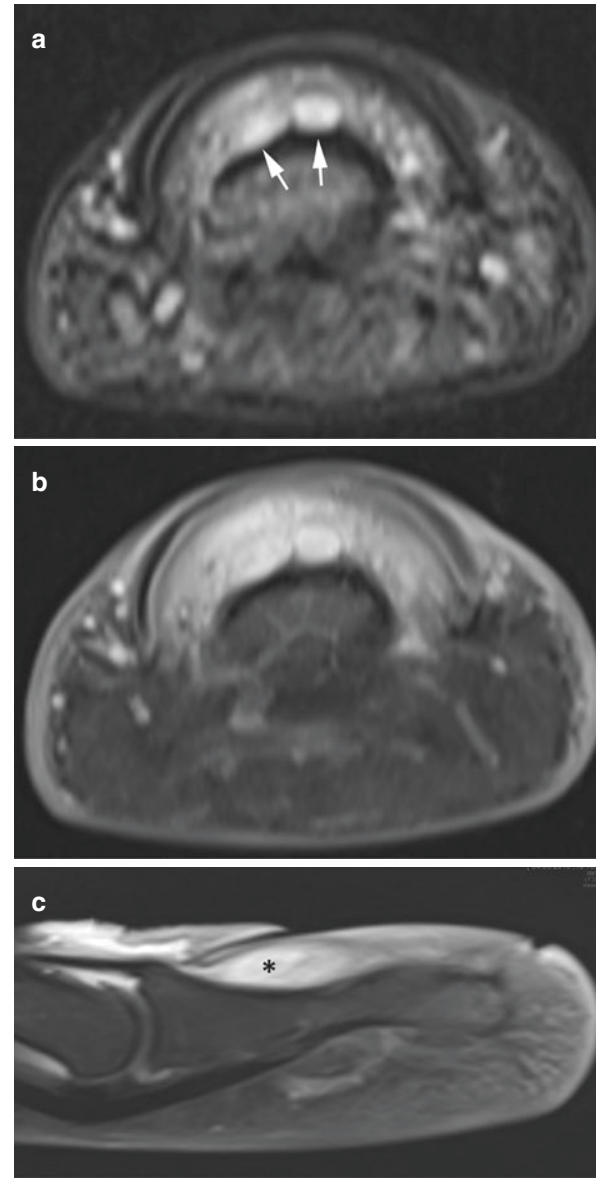

Fig. 4.7 Glomus tumor in the submatrical dermis of the right fourth finger of a 75-year-old female. (**a**) Axial STIR, (**b**) axial, and (**c**) sagittal T1 fat-sat WI after intravenous injection of gadolinium: the bilobulated glomus tumor (arrows, *) is faintly visible due to the high signal of the surrounding submatrical dermis

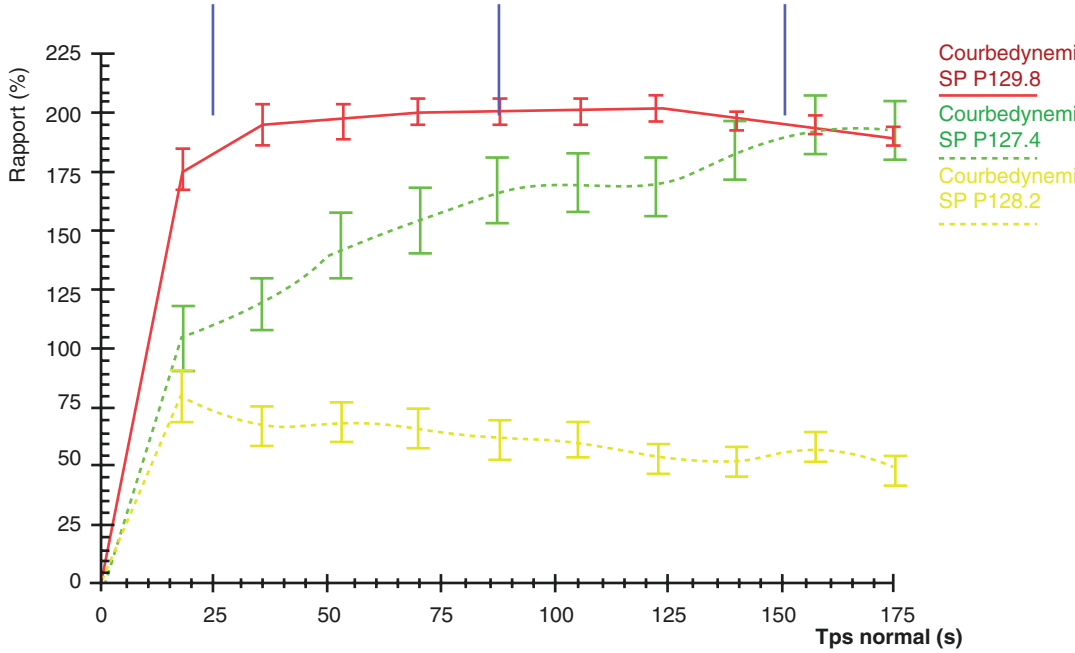

Fig. 4.8 Dynamic enhancement curve of signal after intravenous injection of gadolinium. Red: arterial curve, green: glomus tumor curve, yellow: dermis curve. The glomus tumor presents an early and fast enhancement at the arterial phase and more progressive enhancement on the delayed phase

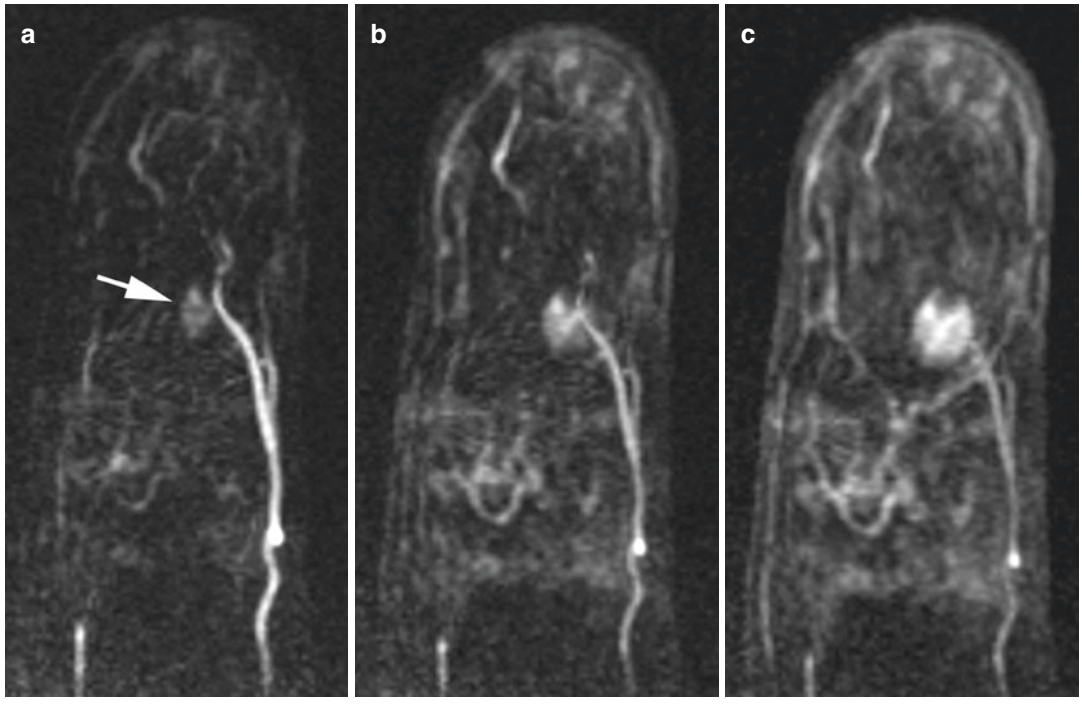

Fig. 4.9 MR angiography of a glomus tumor of the right fourth finger of a 41-year-old female. (**a**) Arterial phase with a dominant radial digital artery and early enhancement of the tumor. Progressive enhancement on the more delayed phases (**b**, **c**)

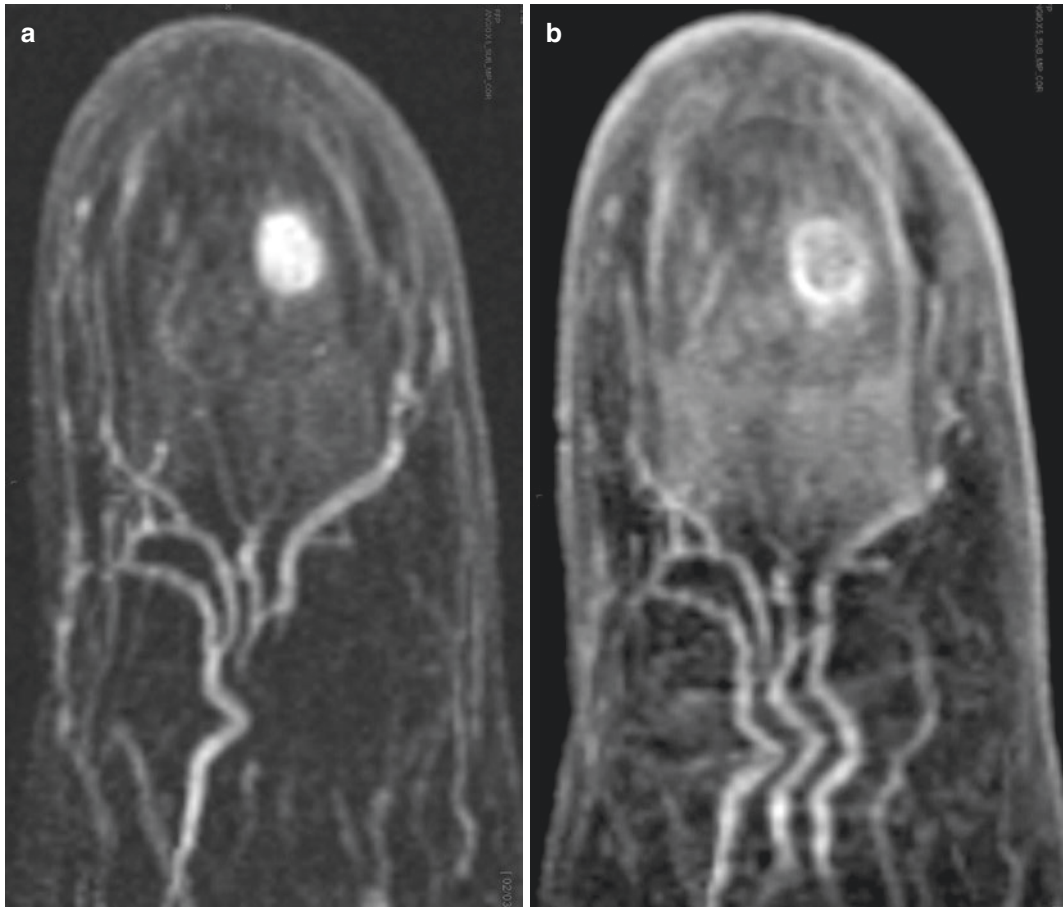

Fig. 4.10 Glomus tumor of the third finger of a 63-year-old female. MR angiography at the (**a**) arterial phase and (**b**) a delayed phase: early and strong enhancement of the tumor. Note the washout of the central part of the tumor on the delayed phase

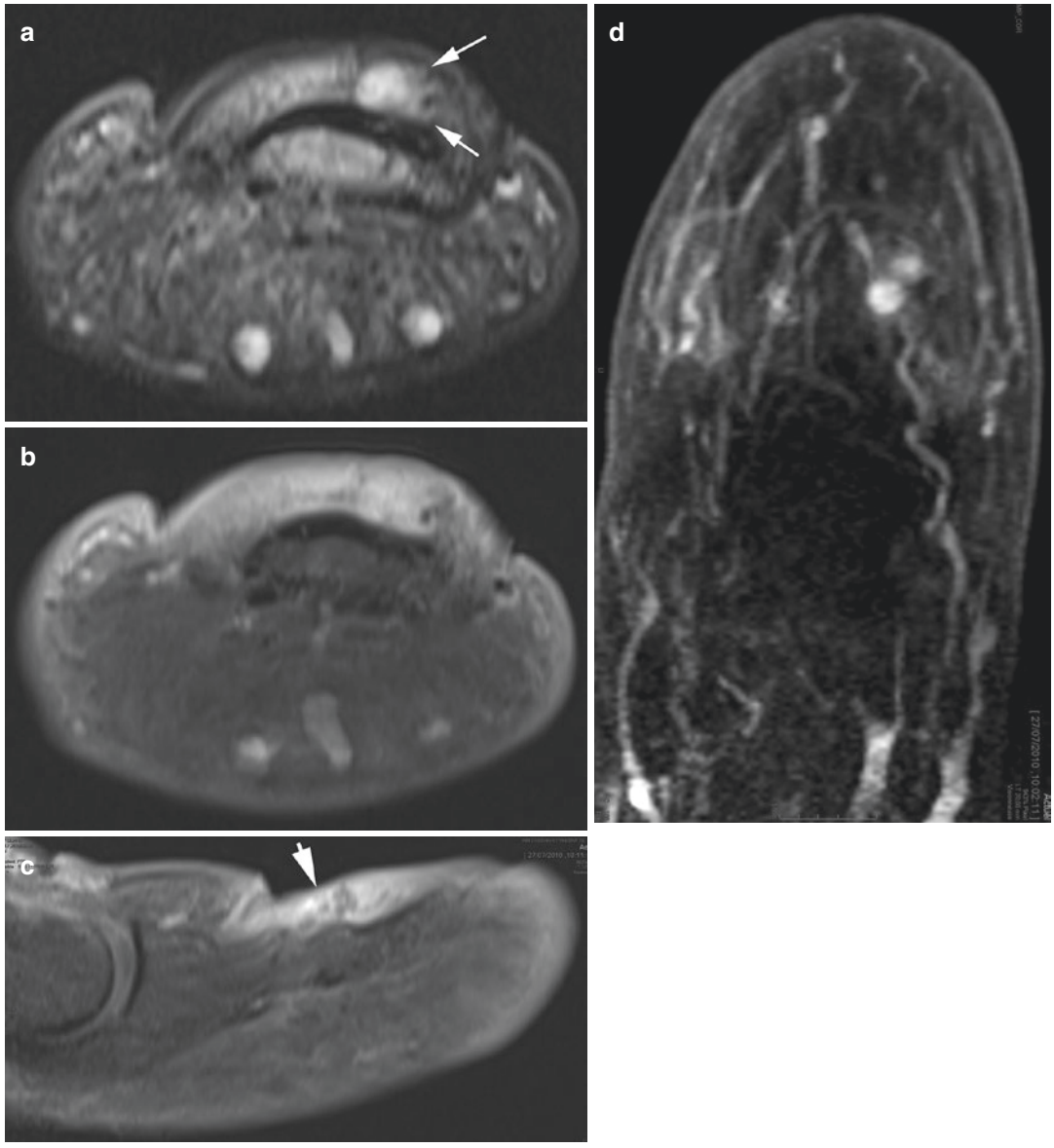

Fig. 4.11 Recurrent glomus tumor of the right thumb of a 63-year-old female. Surgery 5 years before. Recurrent pain after a 6-month free period. (**a**) Axial T2 fat-sat, (**b**) axial, and sagittal (**c**) T1 fat-sat after intravenous injection of gadolinium: the tumor is barely visible with ill-defined limits and postoperative artifacts (arrows). (**d**) MR angiography shows in fact two well-defined contiguous recurrent tumors

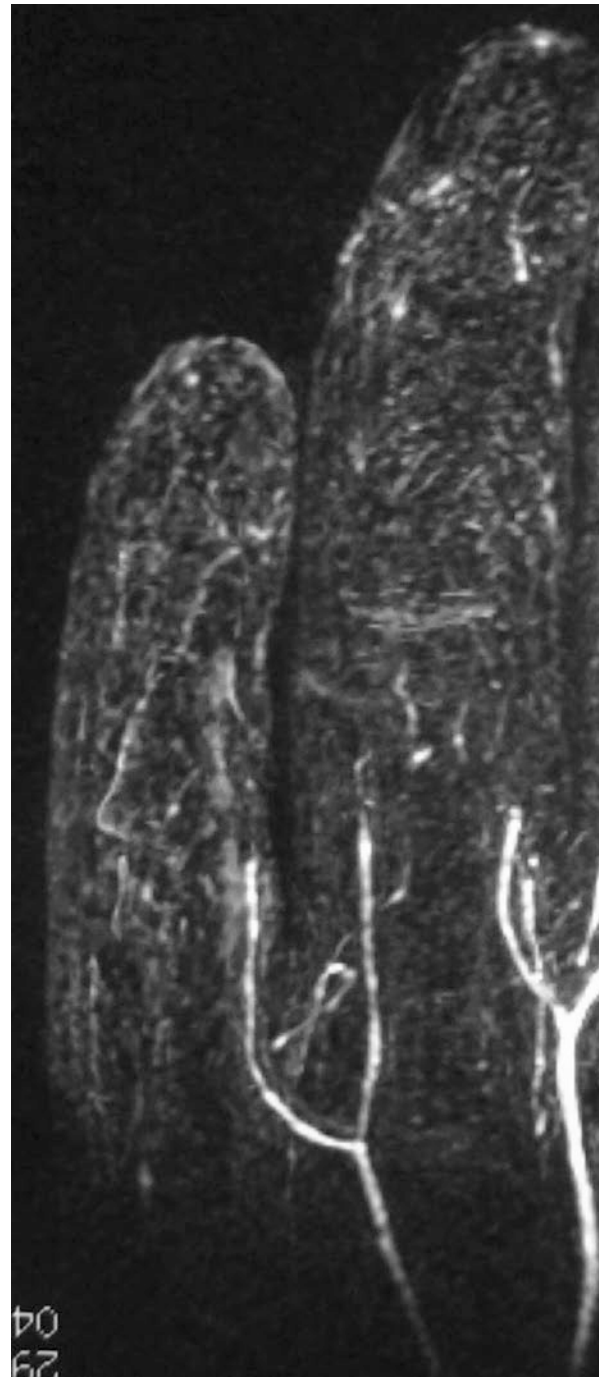

Fig. 4.12 Raynaud's syndrome with a lack of visibility of the vascularization of the fingertips of the fourth and fifth fingers. Note the interrupted thin proximal digital arteries. The detection of the glomus tumor is not possible with MR angiography in this condition

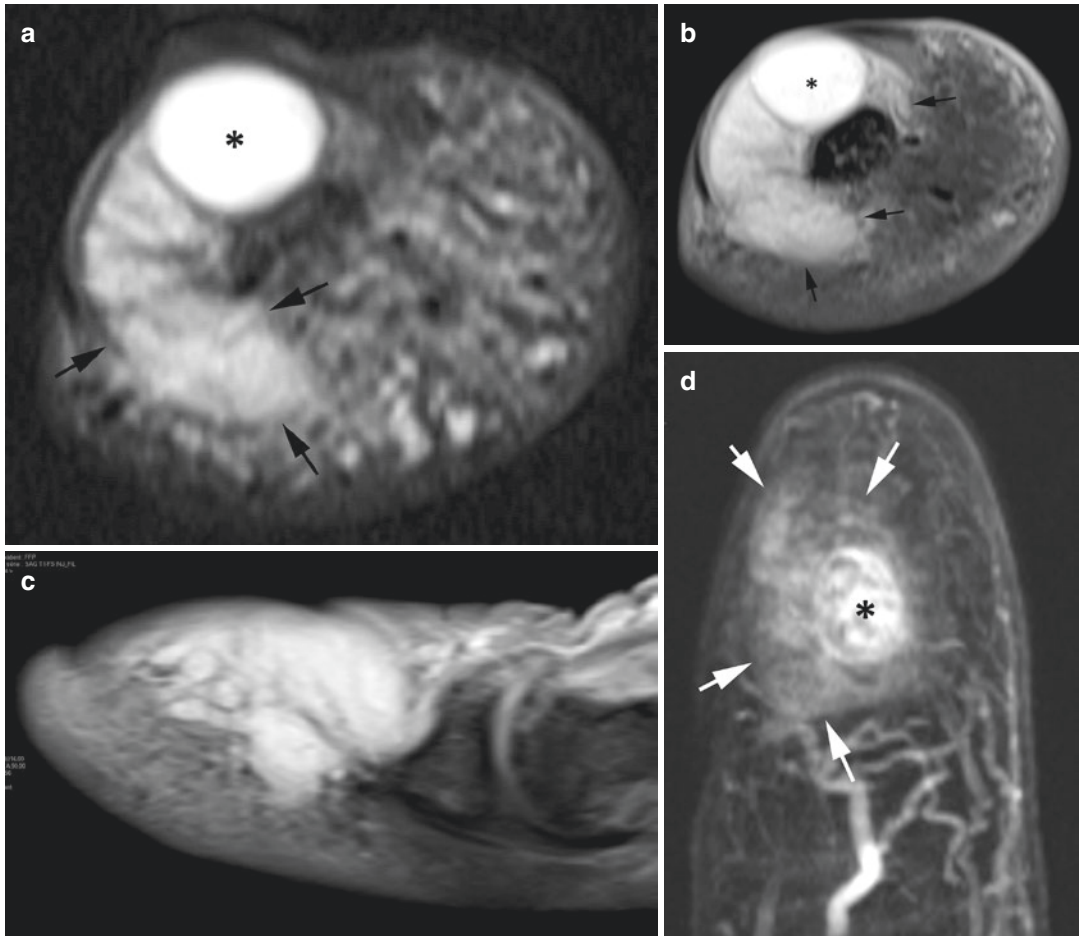

Fig. 4.13 Glomangioma of the left thumb of a 37-year-old female. (**a**) Axial STIR, (**b**) axial, and (**c**) sagittal T1 WI after intravenous injection of gadolinium show an atypical glomus tumor with a round well-defined highly vascularized mass (*) on the midline of the nail bed and infiltrative expansion in the radial part of the nail bed and the pulp (arrows). (**d**) MR angiography better shows these two components of the tumor

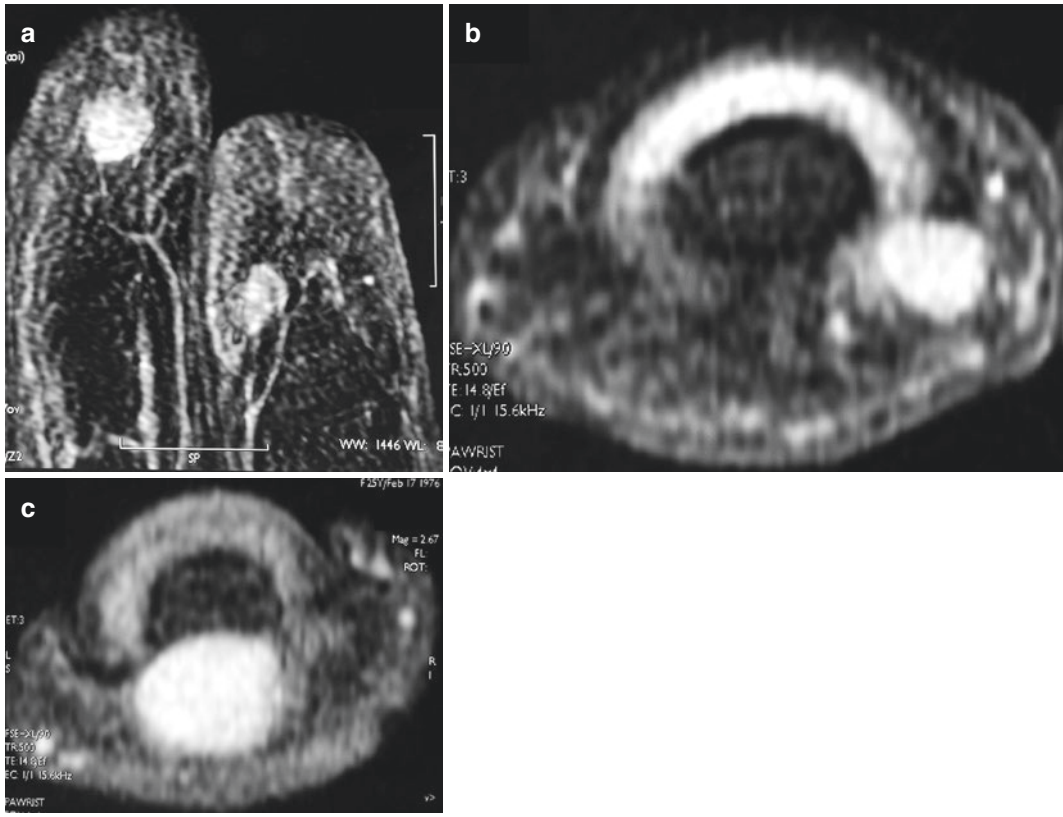

Fig. 4.14 Multiple glomus tumors of the second and third fingers of a 25-year-old female associated with NF1. (**a**) MR angiography shows one glomus tumor on both fingers. Axial STIR (**b**) of the second finger and (**c**) axial T1 fat-sat after intravenous injection of gadolinium of the third (**b**) finger highlight rare locations in the pulp for both fingers

References

1. Chou T, Pan SC, Shieh SJ, Lee JW, Chiu HY, Ho CL. Glomus tumor: twenty-year experience and literature review. Ann Plast Surg. 2016;76(Suppl 1):S35–40.
2. Bargon CA, Mohamadi A, Talaei-Khoei M, Ring DC, Mudgal CS. Factors associated with requesting magnetic resonance imaging during the management of glomus tumors. Arch Bone Joint Surg. 2019;7(5):422–8.
3. Jouari OE, Gallouj S, Elloudi S, Senhaji G, Rimani M, Mermissi FZ. A painless glomus tumor: a case report. J Med Case Rep. 2018;12:302.
4. Kumar MG, Emnett RJ, Bayliss SJ, Gutmann DH. Glomus tumors in individuals with neurofibromatosis type 1. J Cutan Aesthet Surg. 2012;5(4):291–4.
5. Trehan SK, Athanasian EA, DiCarlo EF, Mintz DN, Daluiski A. Characteristics of glomus tumors in the hand not diagnosed on magnetic resonance imaging. J Hand Surg Am. 2015;40(3):542–5.
6. Wong GN, Nandini CL, Teoh LC. Multiple intraneural glomus tumors on a digital nerve: case report. J Hand Surg Am. 2013;38:1972–5.
7. Shinohara T, Hirata H. Glomus tumor originating from a digital nerve. Plast Reconstr Surg Glob Open. 2019;7:e2053.
8. Lu H, Chen LF, Chen Q. Rupture of a subungual glomus tumor of the finger. BMC Cancer. 2018;18:505.
9. Drapé JL, Idy-Peretti I, Goettmann S, Guérin-Surville H, Bittoun J. Standard and high-resolution magnetic resonance imaging of glomus tumors of toes and fingertips. J Am Acad Dermatol. 1996;35(4):550–5.
10. Matloub HS, Muoneke VN, Prevel CD, Sanger JR, Yousif NJ. Glomus tumor imaging: use of MRI for localization of occult lesions. J Hand Surg Am. 1992;17(3):472–5.

11. Al-Qattan MM, Al-Namla A, Al-Thunayan A, Al-Subhi F, El-Shayeb AF. Magnetic resonance imaging in the diagnosis of glomus tumours of the hand. J Hand Surg Br. 2005;30(5):535–40.
12. Ham KW, Yun IS, Tark KC. Glomus tumors: symptom variations and magnetic resonance imaging for diagnosis. Arch Plast Surg. 2013;40(4):392–6.
13. Stewart DR, Sloan JL, Yao L, et al. Diagnosis, management, and complications of glomus tumors of the digits in neurofibrimatois type 1. J Med Genet. 2010;47:525–32.
14. Ofaiche J, Chauvel A, Cogrel O. Solitary subungual glomangioma. J Am Acad Dermatol. 2014;71(1):44–8.
15. Tony G, Hauxwell S, Nair N, Harrison DA, Richards PJ. Large plaque-like glomangioma in a patient with multiple glomus tumours: review of imaging and histology. Clin Exp Dermatol. 2013;38:693–700.