



(CASE REPORT)



IMAGING DIAGNOSIS OF SUBUNGUAL GLOMUS TUMOR: A CASE REPORT

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ABSTRACT

Glomus tumors are rare benign perivascular neoplasms arising from the neuromyoarterial glomus body, a thermoregulatory arteriovenous structure of the reticular dermis. They account for 1-5% of all hand tumors and show a marked predilection for the subungual region of the fingers, with a strong female predominance. Because the classic triad of paroxysmal pain, point tenderness, and cold intolerance is inconstant and the lesion is frequently invisible, diagnosis is commonly delayed by several years. We report the case of a 35-year-old woman, with a history of repeated nail microtrauma related to regular manicures and the use of artificial nails, referred for a three-month history of a painful bluish subungual swelling of the thumb. High-frequency ultrasound demonstrated a well-defined oval nodule of the nail bed that was isoechoic to the adjacent soft tissues and therefore barely conspicuous on B-mode imaging; color Doppler interrogation revealed marked hypervascularity and was the decisive detection tool. Magnetic resonance imaging (MRI) confirmed a small subungual lesion of the distal phalanx, markedly hyperintense on fat-suppressed proton density (PD)/T2-weighted images, hypointense on T1-weighted images, with early, intense, and progressive enhancement on subtracted dynamic contrast-enhanced sequences. The combination of a typical location, marked Doppler hypervascularity, and avid dynamic enhancement was considered diagnostic, and the patient was referred for transungual excision, histological confirmation being pending. This observation illustrates two practical points: in the nail bed, hypervascularity rather than echogenicity is the diagnostic signature of a glomus tumor, and dynamic subtraction markedly increases the conspicuity of a millimetric enhancing nodule.

Keywords: Color Doppler ultrasound; Dynamic contrast-enhanced MRI; Fingertip pain; Glomus tumor.

1 INTRODUCTION

Glomus tumors are benign neoplasms arising from the glomus body, a contractile neuromyoarterial structure of the reticular dermis that regulates cutaneous blood flow and temperature [1-3], and were individualized by Masson in 1924 under the term neuromyoarterial tumor [1]. They account for approximately 1-5% of all tumors of the hand [1] and for

about 2% of all soft-tissue tumors of the extremities [2]; because glomus bodies are most densely distributed in the fingertips, the subungual region of the distal phalanx is by far the most frequent site of occurrence [2,3].

The typical patient is a middle-aged woman: women account for about 71% of reported patients, the mean age at diagnosis is close to 41 years, and the thumb is the digit most frequently involved [3]. The classic triad described by Carroll and Berman, namely spontaneous paroxysmal pain, extreme pain on light pressure, and intolerance to cold, remains the cornerstone of clinical suspicion, reinforced by several bedside tests (Love's pin test, Hildreth's test, the cold sensitivity test, and transillumination) [2,3]. In practice, however, the triad is inconstant and the lesion is frequently invisible, a clinically appreciable nodule being reported in only 17.3% of patients [3]. Patients are therefore commonly misdiagnosed as having a neuroma, a rheumatological disorder, or a chronic nail infection, with a mean delay of 3.9 years between symptom onset and diagnosis [3].

Imaging is consequently decisive. Plain radiographs mainly serve to exclude an underlying bone lesion [1,3], whereas ultrasound and magnetic resonance imaging (MRI) are the two modalities that carry the diagnosis, the latter typically showing a nodule that is hypointense on T1-weighted images, markedly hyperintense on T2-weighted images, and avidly enhancing after gadolinium administration [2-4]. We report the case of a young woman with a subungual glomus tumor of the thumb, illustrating the complementary contribution of high-frequency color Doppler ultrasound and of dynamic contrast-enhanced MRI with subtraction, a technique seldom detailed in the imaging literature devoted to this entity.

2 CASE PRESENTATION

A 35-year-old woman with no relevant past medical history, but reporting repeated minor trauma to the nail apparatus in relation to regular manicures and the recurrent use of artificial nails, was referred to our department for a three-month history of pain of the first digit (thumb) of the right hand. The pain was spontaneous, paroxysmal, and lancinating, markedly exacerbated by minimal pressure on the fingertip and by exposure to cold, and had progressively interfered with manual activities and with sleep. Clinical examination revealed a discrete bluish discoloration and swelling of the nail bed (Figure 1), associated with exquisite, strictly localized tenderness reproduced by the pressure of a pinhead (positive Love's pin test). The nail plate showed no dystrophy, thinning, or longitudinal ridging, and there were no local inflammatory signs. Distal sensation, capillary refill, and interphalangeal mobility were normal, and examination of the remaining digits of both hands was unremarkable.



Figure 1: Clinical photograph of the affected thumb: bluish swelling at the base of the nail of the first digit (arrows).

Ultrasound was performed with a high-frequency linear array transducer (18 MHz) using a thick gel standoff and minimal transducer pressure. On grey-scale imaging, a well-defined oval nodule of the nail bed was identified deep to the nail plate, measuring 7.3×3.4 mm. The nodule was isoechoic to the adjacent nail bed soft tissues and was consequently poorly conspicuous on B-mode imaging alone. Color Doppler interrogation, performed with a low pulse repetition frequency (velocity scale ± 5 cm/s), a low wall filter, and a color gain adjusted just below the noise threshold, demonstrated marked hypervascularity within and around the nodule, and was the decisive finding that allowed the lesion to be confidently localized (Figure 2). The dorsal cortex of the distal phalanx immediately deep to the lesion was regular and continuous.

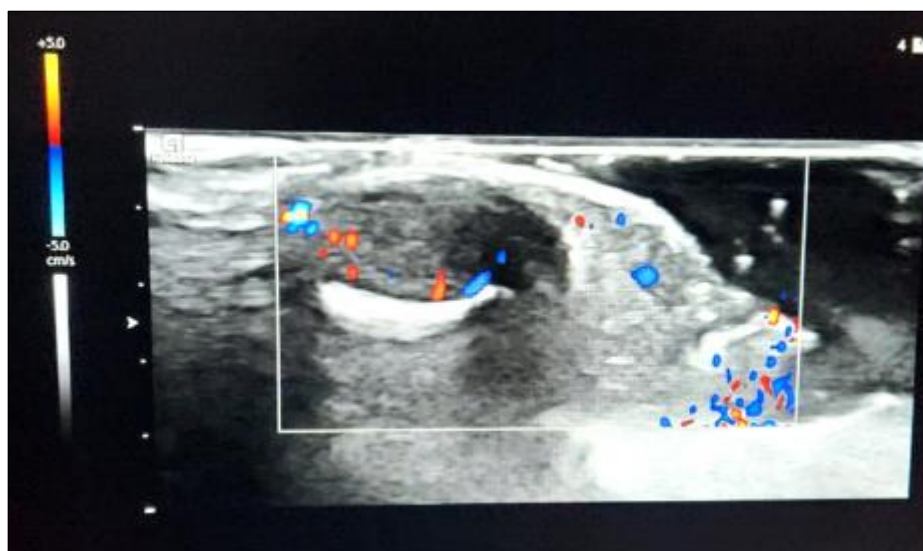


Figure 2: Longitudinal high-frequency color Doppler ultrasound of the distal phalanx: well-defined subungual nodule of the nail bed, isoechoic to the adjacent soft tissues, showing marked vascularity on color Doppler.

MRI of the hand was subsequently performed on a 1.5-T unit with a dedicated surface coil, using thin-section coronal T1-weighted and coronal fat-suppressed PD/T2-weighted sequences, completed by a dynamic contrast-enhanced fat-suppressed T1-weighted acquisition with subtraction after intravenous injection of a gadolinium chelate. A small, well-circumscribed subungual nodule was identified along the ulnar aspect of the distal phalanx of the thumb. The lesion showed marked signal hyperintensity on the fat-suppressed PD/T2-weighted sequence and low to intermediate signal intensity on the T1-weighted sequence (Figure 3). On the subtracted dynamic acquisition, the nodule demonstrated early, intense, and progressively increasing enhancement throughout the dynamic series (Figure 4). There was no cortical erosion of the distal phalanx, no extension towards the germinal matrix, and no additional enhancing nodule elsewhere in the hand or in the other digits.

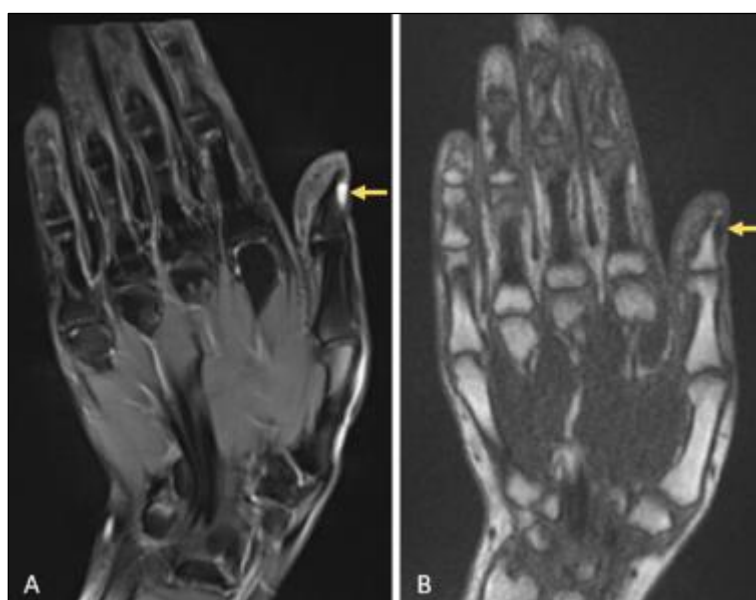


Figure 3: Coronal MRI of the hand. (A) Fat-suppressed PD/T2-weighted image: the subungual lesion of the thumb shows marked signal hyperintensity (arrow). (B) T1-weighted image: the same lesion shows low to intermediate signal intensity (arrow). MRI, magnetic resonance imaging; PD, proton density

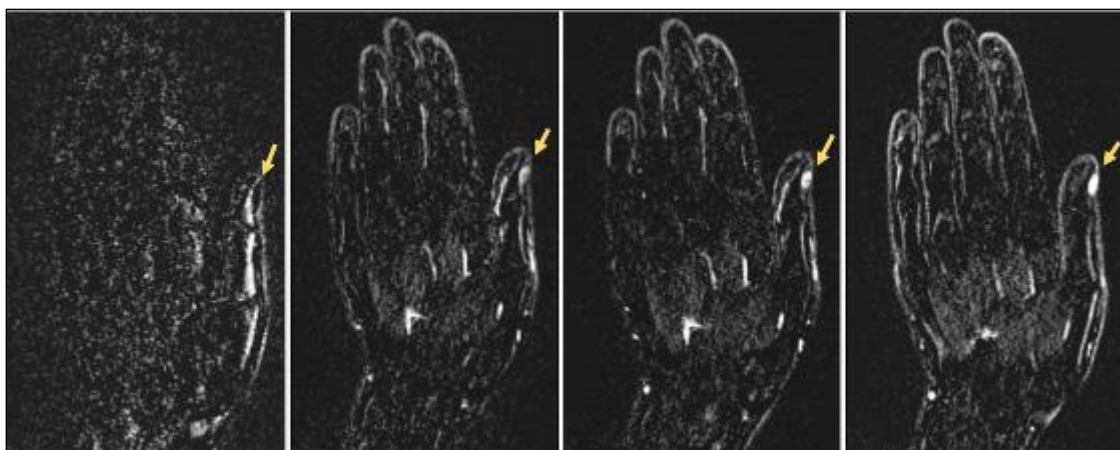


Figure 4: Subtracted dynamic contrast-enhanced MRI of the hand. Successive dynamic phases from left to right: the subungual lesion of the thumb shows early, intense, and progressive enhancement (arrows). *MRI, magnetic resonance imaging*

The combination of a typical clinical presentation, a hypervascular subungual nodule on color Doppler ultrasound, and a characteristic MRI signal and enhancement pattern was considered diagnostic of a glomus tumor. The patient was referred to the hand surgery unit for complete excision through a transungual approach. At the time of writing, surgery was pending.

3 DISCUSSION

Our patient reproduces almost point by point the epidemiological profile reported in the literature. In the narrative review by Pietramala et al., which pooled 1142 patients from 131 studies, 815 (71.3%) were women, the mean age at diagnosis was 40.8 years, and the thumb was the digit most frequently involved [3]. In the 20-year single-institution experience reported by Chou et al., the female-to-male ratio was 3:2 overall and 4:1 in the digital subgroup, the mean age was 42.3 years, 66% of the tumors were digital, and 32 of the 33 digital tumors were subungual, with a mean size of 4.3 mm in the digital subgroup [2]. A Moroccan series of 11 patients published from our own institution reported nine women and two men with a mean age of 36.3 years (range 24–54 years) and tumors measuring 4 to 10 mm [1]. A 35-year-old woman with a subungual tumor of the thumb therefore corresponds precisely to the modal presentation of the disease.

The etiology of glomus tumors remains incompletely elucidated, but sex, age, trauma, and inheritance have all been implicated in their growth, repeated traumatic episodes being thought to induce a reactive hypertrophy of the glomus body through a weak point in its structure [3]; a history of trauma to the affected area has been specifically reported in association with subungual glomus tumors [2]. Our patient reported repeated minor trauma to the nail apparatus related to regular manicures and to the recurrent application of artificial nails. A single observation obviously cannot establish causality, but this increasingly widespread cosmetic exposure deserves to be mentioned for two reasons. First, it constitutes a plausible source of the repeated microtrauma incriminated in the literature, and it should therefore be actively sought when taking the history of a patient with chronic fingertip pain. Second, it is a powerful source of diagnostic confusion, since such patients are readily labelled as having a chronic nail disorder and treated as such, a scenario that contributes to the diagnostic delay characteristic of this tumor [3].

The classic triad is far less constant than its reputation suggests. In the pooled review, pain was present in 76.2% of the patients for whom the information was available (182/239), cold sensitivity in 47.3% (113/239), a clinically visible mass or nodule in only 17.3% (47/272), and nail deformity in 22.7% (54/238) [3]. Chou et al. reported pain and tenderness in 90% of their 50 patients, and in 100% of the digital subgroup, but cold hypersensitivity in only 30% [2]. In the Moroccan series, all 11 patients were painful, with a mean visual analogue scale score of 6/10 [1]. Bedside tests are helpful but imperfect: the cold sensitivity test has been credited with 100% sensitivity, specificity, and accuracy, Love's pin test with 100% sensitivity but only 78% accuracy, and Hildreth's test with 100% specificity, 71.4% sensitivity, and 78% accuracy, which is why a combination of tests rather than any single test is recommended [2,3]. Even so, clinicians cannot rely on physical examination alone [3], and the differential diagnosis remains broad, including leiomyoma, hemangioma, neuroma, and gouty arthritis for solitary lesions, blue rubber bleb nevus syndrome for multiple lesions [3], and fibroma, neuralgia, or inflammatory arthropathy [1]. The practical consequence is a diagnostic delay averaging 3.9 years in a series of 75 patients [3]. In this respect, our patient was diagnosed unusually early, three months after symptom onset, most probably because the bluish subungual discoloration, a sign present in only a minority of patients, was recognized and led directly to dedicated imaging.

Radiographs were systematically obtained and were normal in all 11 patients of the Moroccan series, their main value being the exclusion of an underlying bone lesion [1]. Pietramala et al. similarly observed that radiographs are commonly requested but usually add no information about the pain or the mass, although cortical thinning or erosive changes of the adjacent bone may be seen in some cases [3].

Ultrasound is inexpensive, non-invasive, widely available, and free of contraindication, and it typically depicts a hypoechoic lesion with clear boundaries and a regular shape, often accompanied by internal flow signals [3]. Its performance has improved considerably with high-frequency probes, which now resolve the individual structures of the nail unit and characterize tumors, cysts, and vascular malformations [3]; subungual glomus tumors smaller than 2 mm have been localized in this way [3], and color Doppler sonography has been credited with a 100% detection rate in digital glomus tumors [3], color duplex Doppler showing no false-negative result in the series of Chen et al. [2]. On the strength of these data, Chou et al. proposed radiography and Doppler sonography as the first-line imaging tools for subcutaneous lesions and considered MRI a second-line examination reserved for multiple or recurrent tumors, on account of a comparable sensitivity, a lower specificity, and a higher cost [2].

The limitations of ultrasound are equally well identified: it is operator-dependent and requires both a radiologist trained in musculoskeletal imaging and an adapted transducer [1]; the curvature of the nail plate may generate artifacts in the lateral nail folds; and small, flattened subungual lesions may escape detection [3]. Our observation adds a practical nuance to this description. The nodule was not hypoechoic but isoechoic to the surrounding nail bed, and was consequently almost invisible on grey-scale imaging alone: detection rested entirely on the color Doppler signal. This underlines that in the nail bed the diagnostic information is carried by vascularity rather than by echogenicity, and that the Doppler settings matter as much as the probe frequency. A thick gel standoff and minimal transducer pressure are essential, since even light compression of the fingertip may obliterate flow within these small, low-velocity vessels; the pulse repetition frequency, the wall filter, and the color gain must be optimized for slow flow, a velocity scale of ± 5 cm/s being used in the present case. A technically sub-optimal Doppler examination may therefore generate a false-negative result in a lesion that is, by nature, hypervascular.

MRI is the most frequently employed modality in the published literature, having been performed in 509 of 1142 patients (44.6%), ahead of ultrasound (299 patients) and radiographs (229 patients), whereas angiography was used exceptionally [3]. The signal pattern is well established and was faithfully reproduced in our patient: low signal intensity on T1-weighted images, marked hyperintensity on T2-weighted images, and intense enhancement after gadolinium injection [2-4]. A high-signal central dot surrounded by a rim of lower signal intensity, the so-called nidus appearance, is regarded as more specific but is inconstant [2,3]. It must be remembered that this signal pattern is shared by any hypervascular soft-tissue lesion, so that it is only in a typical location that it should raise the suspicion of a glomus tumor [3].

The diagnostic performance of MRI is markedly asymmetrical. In a series of 42 patients, sensitivity reached 90% and the positive predictive value 97%, so that a positive MRI in a clinically suspected glomus tumor is highly suggestive of the diagnosis; specificity, however, was only 50% and the negative predictive value as low as 20%, so that a negative examination does not exclude the lesion [2,3], MRI having been reported to yield negative results in up to 10% of cases [3]. Part of this limitation is technical: thresholds of 3 mm and of 1.5 mm have both been proposed for the smallest detectable lesion, and Drapé et al. showed that tumors as small as 2 mm could be detected only with a dedicated local surface gradient coil (1.5-cm radius, 117-mm pixel size) requiring modification of the 1.5-T unit [3]. Since the average subungual glomus tumor measures about 6 mm (range 1.9-10 mm), standard protocols are adequate for most lesions but will miss the smallest ones [3].

It is precisely here that the dynamic contrast-enhanced acquisition with subtraction used in our patient appears valuable. Subtraction cancels the intrinsically high T1 signal of the surrounding fat and of any hemorrhagic component, so that a millimetric enhancing nodule stands out against a uniformly suppressed background, as illustrated in Figure 4. In addition, the temporal behavior of the enhancement, which was early, intense, and progressive, provides a functional argument that corroborates the hypervascularity demonstrated by color Doppler, whereas a single post-contrast acquisition documents only the presence of enhancement. Finally, because the acquisition covered the entire hand, synchronous lesions could be excluded during the same examination, an issue that is not trivial given that multiple forms exist and may be mistaken for blue rubber bleb nevus syndrome [3], and that the surgical approach must be designed so as to address synchronous or metachronous lesions [2].

Arteriography, and more specifically MR angiography, can display the vascular component of the tumor as a telangiectatic vascular lake, an appearance considered pathognomonic; it is, however, invasive and expensive, has been used in only a very small minority of the reported patients, and is now reserved for atypical or inconclusive cases [3]. Computed tomography is used only rarely and mainly in association with other modalities [3]. The trend reported in the recent literature is a progressive shift towards less invasive and more accessible imaging, with MRI as the reference

standard and high-frequency ultrasound with color Doppler emerging as a cost-effective alternative [3], a trend that our observation illustrates in practice.

Chou et al. proposed a simple algorithm in which classic symptoms lead to the three clinical tests, then to confirmation by MRI or Doppler sonography, and finally to histopathological diagnosis [2]. Our observation allows this pathway to be refined from the radiologist's standpoint. In a patient with a painful subungual lesion in a typical location, a meticulously performed high-frequency color Doppler ultrasound is a reasonable first-line examination: it is inexpensive, immediately available, free of contraindication, and its spatial resolution within the nail unit equals or exceeds that of a standard MRI protocol. MRI, ideally including a dynamic contrast-enhanced acquisition with subtraction, should be obtained when ultrasound is negative or equivocal despite strong clinical suspicion, when the location is atypical, when multiple lesions are suspected, in the setting of recurrence, or whenever precise preoperative mapping is required. Neither modality replaces histopathology, which remains the only means of definitive confirmation [1].

Complete surgical excision is the only effective treatment, and incomplete excision is the main cause of recurrence [3]. The transungual approach, used in nine of the 11 patients of the Moroccan series [1] and in all 33 subungual tumors of Chou et al. [2], provides optimal exposure of the nail bed; postoperative nail deformity occurred in 4% of patients in the latter series, with no recurrence [2], and in two of 11 patients in the former, without functional or aesthetic consequence [1]. The lateral subperiosteal approach spares the nail and is particularly suited to proximal subungual lesions close to the germinal matrix, at the cost of restricted visibility and a higher probability of incomplete excision [1,2]. Reported recurrence rates vary widely and may reach 50% in some series; early recurrence generally reflects incomplete excision or a second tumor missed at diagnosis, whereas late recurrence usually corresponds to a genuinely new lesion [3]. This distinction gives the preoperative imaging assessment a direct therapeutic relevance and justifies repeating imaging in case of recurrence, in order to exclude a new or a malignant lesion [2]. Malignant transformation is exceptional [1]; the criteria for malignancy are a deep location, a size greater than 2 cm, atypical mitotic figures or marked nuclear atypia, and five or more mitoses per 50 high-power fields [2]. None of these features was present in our patient.

This report carries the limitations inherent to a single observation. Plain radiographs, classically obtained as a first-line examination [1,3], were not performed, ultrasound having been carried out directly at the time of referral. Ultrasound was performed with a standard high-frequency musculoskeletal transducer and MRI with a standard hand protocol, rather than with the dedicated micro-coil technique described for millimetric lesions [3]; the excellent lesion conspicuity obtained here is therefore explained by the size of the tumor and by the dynamic subtraction technique rather than by dedicated hardware. Quantitative perfusion analysis of the dynamic series was not performed, and the prolonged follow-up required to exclude late recurrence is not yet available. The causal role of the repeated cosmetic nail trauma reported by our patient cannot be established from a single case. Above all, surgical excision had not been performed at the time of writing, so that the diagnosis rests on the concordance of the clinical presentation with two independent imaging modalities and still awaits histopathological confirmation, which remains the reference standard [1].

4 CONCLUSION

Subungual glomus tumor should be considered in any patient with chronic, strictly localized fingertip pain aggravated by pressure and by cold, and all the more so when a bluish subungual discoloration is present, a history of repeated nail trauma being an additional argument that should be actively sought. The present case illustrates a diagnostic pathway that is reproducible in daily practice: a meticulously performed high-frequency color Doppler ultrasound identified a nodule that was isoechoic and therefore almost invisible on grey-scale imaging, and a dynamic contrast-enhanced MRI with subtraction confirmed the diagnosis, defined the exact relationship of the lesion with the nail matrix and the distal phalanx, and excluded synchronous lesions. In the nail bed, hypervascularity rather than echogenicity is the diagnostic signature of the glomus tumor, and the technical quality of the Doppler and dynamic acquisitions is the main determinant of diagnostic success. Recognizing this pattern shortens a diagnostic delay that still averages several years and leads directly to the only curative treatment, namely complete surgical excision.

STATEMENTS ON COMPLIANCE WITH ETHICAL STANDARDS

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Disclosure of conflict of interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper. No funding was received.

Statement of ethical approval

This single-patient case report was conducted in accordance with the ethical standards of our institution and with the Declaration of Helsinki. Institutional review board approval was waived for this retrospective, anonymized single-case report.

Statement of informed consent

Written informed consent was obtained from the patient for the publication of this case report and of the accompanying images, all of which were anonymized.

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