
SUBUNGUAL SOLITARY GLOMUS TUMOR

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Abstract

The glomus tumor is a rare, usually solitary tumor, which is difficult to diagnose because of obscure symptoms and the small size of the lesion. Most cases involve subungual sites. The diagnosis is usually made on clinical and histologic grounds, but plain radiographs, ultrasonography and dermoscopy may prove useful. The treatment of choice for symptomatic solitary glomus tumors, despite nail disfigurement, is total surgical excision and the prospects for complete resolution of symptoms are good if excision is complete.

Keywords:

Glomus tumor, subungual, dermoscopy.

INTRODUCTION

Glomus tumors are rare, usually solitary, soft tissue neoplasms that typically present in adults of 30-50 years of age, as small, blue-red papules or nodules of the distal extremities, with most cases involving subungual sites. First described by Hoyer in 1877, these tumors account for less than 2% of all soft tissue tumors and are typically painful, often causing paroxysmal pain in response to temperature changes or pressure. They are thought to arise from the glomus body, a thermoregulatory shunt located in the stratum reticulare throughout the body, but more concentrated in the digits and particularly beneath the nail. Glomus tumors in general show no sex predilection, however, solitary subungual lesions are more commonly observed in women.

The glomus tumor is difficult to diagnose because of obscure symptoms, such as chronic pain and hypersensitivity, and the small size of the lesion. Typical time from symptoms onset to the correct diagnosis is seven years. The initiating event for glomus cell proliferation in glomus tumors, is unknown. Some authors have postulated that trauma induces solitary subungual glomus tumors, although this theory is not well studied.

Histologically, glomus tumors are made up of an afferent arteriole, anastomotic vessel, and collecting venule. They are thought to represent hamartomatous proliferations of modified smooth muscle cells originating from preexisting normal glomus cell populations. Upon biopsy, the histopathology typically reveals organoid appearance of polyhedral cells. It also can display darkly staining nuclei with fibrous stroma, and few blood cells. The lesions contain endothelium-lined vascular spaces surrounded by clusters of glomus cells and they appear mostly as solid, well-circumscribed nodules surrounded by a rim of fibrous tissue.

While the vast majority of glomus tumors are benign, malignant cases have been rarely reported, with such cases typically being locally invasive, and with metastases being exceedingly rare. Criteria for the diagnosis of malignancy in glomus tumors include tumor size of more than 2 centimeters and subfascial or visceral location, rapid growth, deep soft tissue involvement, atypical mitotic figures, marked nuclear atypia and any level of mitotic activity.

The patient with glomus tumor seeks medical attention early, but this can be a difficult diagnosis to make as the mass is frequently too small to be identified on physical examination. Patients classically present with paroxysmal pain, often precipitated by cold or pressure and they may avoid using the affected digit. Nevertheless, the classic triad of paroxysmal pain, temperature sensitivity, and focal tenderness is nonspecific and not all three may be present. The tumors tend to have a bluish discoloration, although a whitish appearance may also be noted. Elevation of the nail bed can occur as the tumour proliferates within the enclosed space. In rare cases, the tumors may present in other body areas, such as the gastric antrum or glans penis.

The diagnosis is usually made on the basis of the clinical history and examination. Two useful findings for diagnosing solitary glomus tumors, are the following: 1) Love test - Eliciting exquisite localized pain by applying pressure to the suspected areas with a pencil tip or pinhead; 2) Hildreth sign - Reduction of pain and tenderness and reduction of tenderness with the Love test by inducing transient ischemia with a tourniquet.

While the diagnosis of a glomus tumor is often made on clinical and histologic grounds, imaging may prove useful, especially in cases in which the diagnosis is in doubt. Plain radiographs may show “scalloping” of the distal phalanx; however, the absence of changes on plain radiographs does not exclude a glomus tumour. Further imaging may show the tumour. Ultrasonography, especially Duplex ultra-sonography has a high detection rate and can detect glomus tumors as small as 2 mm. Magnetic resonance imaging can also be useful.

A growing interest in the diagnosis and treatment of nail disorders transformed an area previously neglected by physicians into an attractive and necessary field of knowledge for dermatologists and resulted in the rapid progress of onychoscopy in the development of criterias for a well-founded assessment of nail diseases. Longitudinal erythronychia, triangular onycholysis and blue spots are observed in nail glomus tumor.

The differential diagnoses should include local infection or osteomyelitis, osteoid osteoma, painful conditions of the nail, malignancy and inclusion cysts, blue nevi, Blue Rubber Bleb Nevus Syndrome, eccrine spiradenoma, Kaposi Sarcoma, leiomyoma, Maffucci Syndrome, neurilemoma, venous malformations.

The treatment of choice for symptomatic solitary glomus tumors is total surgical excision and the prospects for complete resolution of symptoms are good if excision is complete. While various treatment modalities have been reported, to include laser and sclerotherapy, in the case of solitary glomus tumors, complete removal of the tumor capsule is recommended to relieve pain and minimize risk for recurrence. Recurrence is unusual, with a higher incidence in glomus tumors that are skin-colored or located in the nail matrix and is most likely related to incomplete excision or undetected multiple lesions.

CASE REPORT

A 56 years old male patient presented to our clinic complaining about change in color on one fingernail and moderate pain. He referred that the symptoms started a few months ago. The patient did not refer any trauma of the nail or any personal or family history for melanoma.

The microscopic examination and the culture for fungal infection were negative. The X-ray of the distal phalanx showed no bone involvement. Dermatoscopic examination revealed triangular onycholysis and blue spots.

A 6- mm punch biopsy specimen of the matrix and the proximal part of the nail was sent for histopathologic examination. Histopathology revealed organoid appearance of polyhedral cells, endothelium-lined vascular spaces surrounded by clusters of glomus cells. The findings were consistent with a diagnosis of glomus tumor. Complete surgical excision of the tumour was performed.

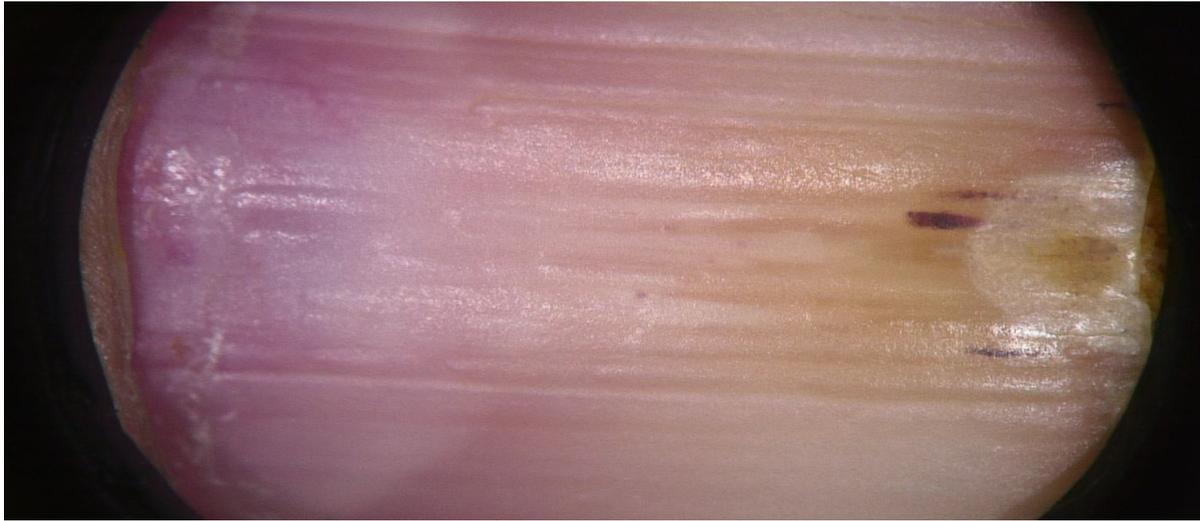


Fig. 1. Dermoscopic view

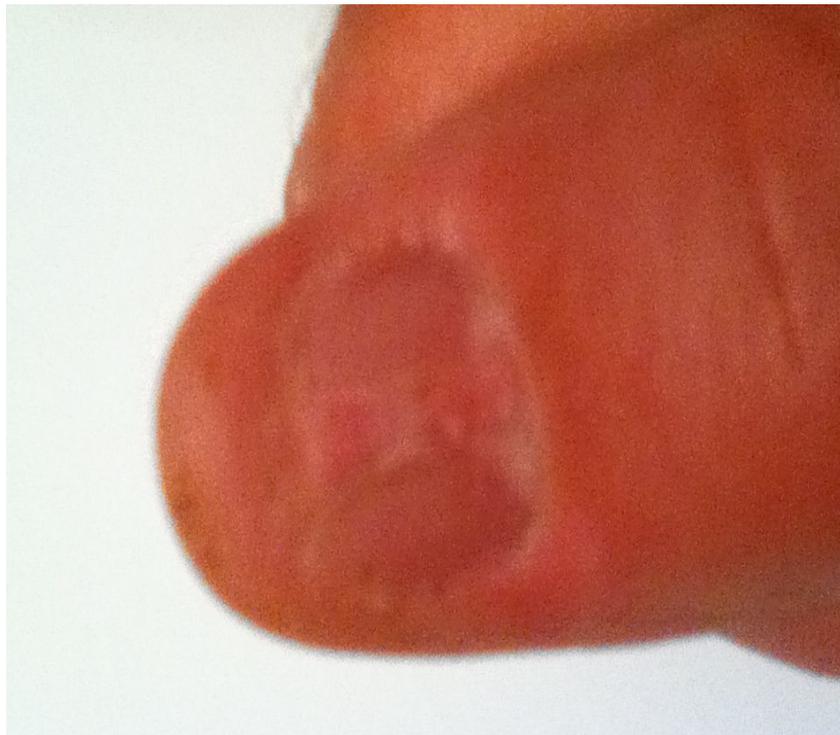


Fig. 2. Post-surgery view

DISCUSSION

Glomus tumor is a benign condition in which a complete excision usually leads to cure, with low incidence of recurrence. However, this benign condition has an unusually high morbidity to the patient before the correct diagnosis is made. This attests to the difficulty in correctly diagnosing this lesion initially. Although history and carefully performed physical examination significantly narrow the differential diagnosis, the plain radiographs are minimally helpful until the bony erosion occurs at the later stages of the disease.

CONCLUSION

With increased index of suspicion, carefully performed history and physical examination, along with the findings on the MRI and dermoscopy, the pre-operative morbidity to the patient with glomus tumor, can be significantly reduced.

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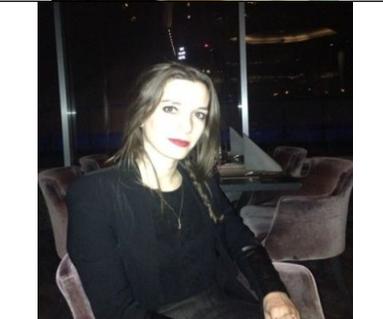
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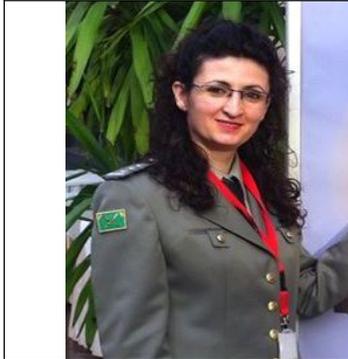


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