Glomus tumor with an atypical location

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ABSTRACT

Background: Glomus tumors are benign and rare vascular tumors, with an incidence of less than 2% in relation to all soft tissue tumors. About 75% occur in hand, especially in the nail bed, even though these tumors may also occur in atypical locations. They are characterized by a triad composed of pain, cold intolerance, and point tenderness.

Case Presentation: We present a clinical case of a male patient with 67 years old with a painful lesion on the subcutaneous tissue of the knee with 5 years of evolution. The pain exacerbated with the cold. He underwent surgical excision with pain resolution, and no recurrence or metastasis was found at 30-month follow-up.

Conclusion: The rapid diagnosis of glomus tumors is important to avoid treatment delays leading to chronic pain. It is important to consider glomus tumors in the differential diagnosis of subcutaneous lesions.

Keywords: Glomus tumor, glomangioma, atypical location, knee, oncology, case report.

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Background

Glomus tumors are rare benign hamartomas, representing less than 2% of all soft tissue tumors [1]. Of these, about 75% occur in the hand [2], and therefore, extradigital tumors are very rare [3]. They originate in the glomus apparatus [3], which is in subcutaneous cellular tissue. The glomus apparatus consists of enlarged vascular channels surrounded by nerve cells and is crucial in temperature regulation. Their malignancy is extremely rare [4].

The tumors are characterized by a classic triad of symptoms composed of pain, cold intolerance, and local tenderness [5]. The tumors occur in adults aged between the fourth and seventh decades of life, with no gender predominance, except those located in the subungual bed that occur more often in females [6].

We report an exceptional case of an atypically located glomus tumor.

Case Presentation

A 67-year-old male, with a prior history of a direct trauma after a fall onto the knee, had a subcutaneous small, painful, and red nodule with a size of 1cm, on the anteriomedial side of the left knee. The Love and Tinel tests were positive. Even though the lesion had 5 years of evolution, there was no noticeable change of the nodule size along time, and the knee's function was unaffected with the full range of motion.

The knee ultrasound revealed a hypoechoic nodule with 12 × 5 mm with unspecified characteristics (Figure 1). The magnetic resonance imaging showed no details of the lesion and a grade IV chondromalacia of the knee on the outerbridge classification.

Intraoperatively, we observed a 12 × 10 × 10 mm red tumor with elastic consistency and well-defined limits, without any adhesions or pedicle in the subcutaneous tissue of the knee (Figures 2 and 3). Marginal excision of the mass was performed, without rupture of the lesion.

A histopathological study identified a well-defined, capsule-free nodular formation consisting of proliferation of blood vessels, and some of them enlarged, branched, lined with low endothelial cells, sometimes containing proteinaceous material such as plasma or red blood cells (Figure 4). At the periphery of the tumor, a thick fibrous septa could be observed with the proliferation of cells with scarce cytoplasm, with round-to-oval, regular, and uniform nuclei without cytological atypia and without significant mitotic activity (Figure 5). Immunohistochemical analysis was negative for SE1/AE3, protein S100, and CD34 (which highlights the blood vessels). CD31 highlights the endothelium of blood vessels present in the lesion. The cells around the blood vessels show positivity for smooth muscle actin, compatible with glomus tumor.

The patient had pain relieved immediately after the surgery. A postoperative wound healing had occurred without complications, and the patient maintains a full range of

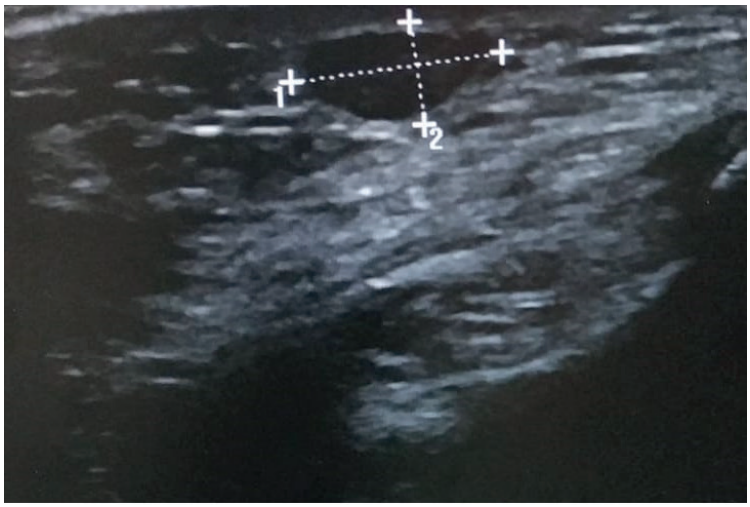

Figure 1. *Ultrasound of the lesion.*

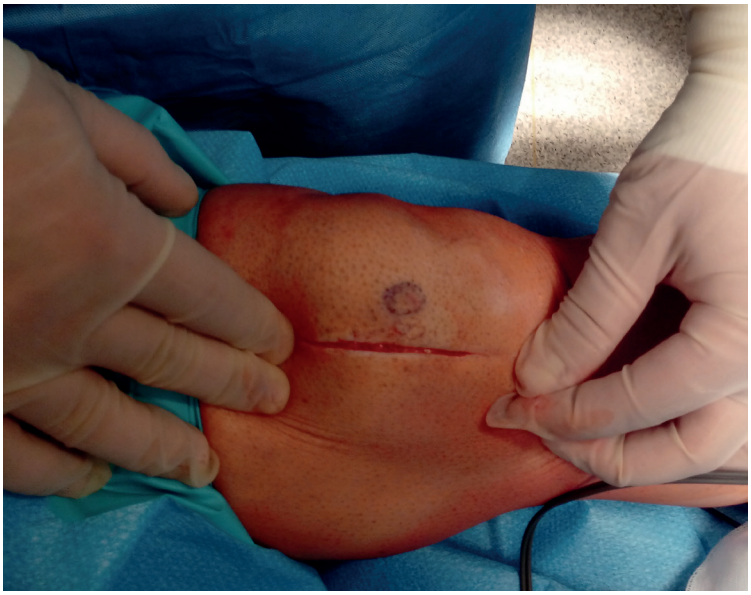

Figure 2. *Surgical approach.*

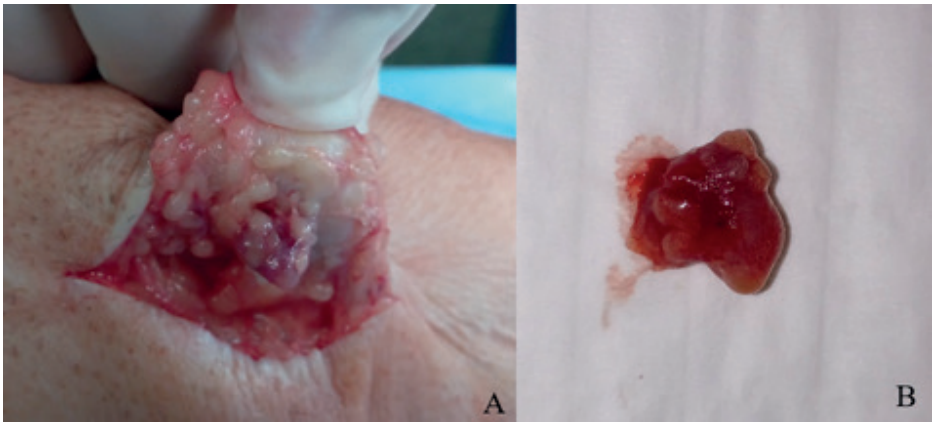

Figure 3. *Intraoperative view of cystic tumor (A); macroscopic aspect of the tumor (B).*

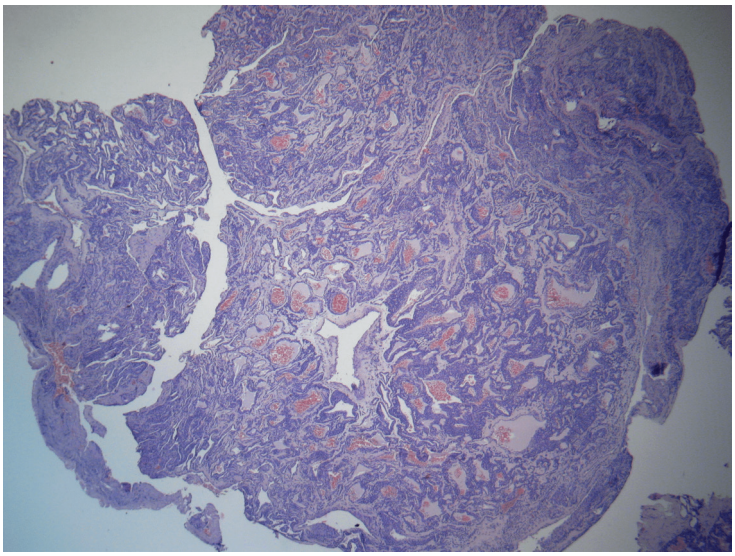

Figure 4. *Well-delimited lesion, of vascular nature, H&E 40×.*

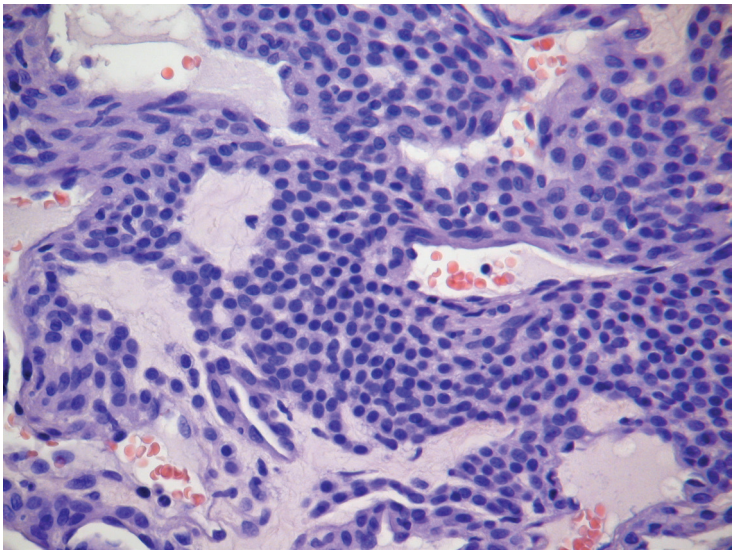

Figure 5. *Between the blood vessels, there is a proliferation of small cells, with scanty cytoplasm and monotonous nuclei, H&E 200×.*

motion of the knee. After 2½ years of follow-up, he presented a total recovery with no pain and no tumor relapse.

Discussion

We present a rare glomus tumor located in the knee of the patient who had the classic triad of symptoms. Ultrasound examination revealed the location and size of the lesion. This prompted us to offer surgical resection as the treatment of choice for this patient. Once the tumor was removed, the diagnosis was confirmed by the histological examination of the resection specimen, and the patient's follow-up for as long as 2½ years revealed no symptoms, disabilities, or tumor relapse.

The normal glomus body is a specialized neuromyoarterial receptor with an afferent arteriole and an efferent venule, a Sucquet-Hoyer anastomotic canal, glomus cells containing actin around the canal, an intraglomerular retinaculum, and a capsule [7]. Its normal function is to regulate temperature and blood pressure, controlling peripheral blood flow [8]. The tumor results from hyperplasia of one or more parts of this glomus body. The cells that made up of the glomus body are characterized by being round or oval. They are specialized perivascular muscle cells and have a dense and granular cytoplasm. Nonmyelinated nerve fibers are responsible for the excruciating pain [7].

Histologically, these tumors can be differentiated into three types: solid, glomangioma, or glomangiomyoma, depending on the predominant component. The first one is a subtype characterized by poor vasculature and scarce smooth muscle. The second is the angiomatoid (glomangioma) with a prevalent vascular component, which was found in the

present clinical case. The last one is the glomangiomyoma, with prominent vascular and smooth muscle components.

Clinically, they are characterized by pain, cold intolerance, and point tenderness [5]. On physical examination, excruciating pain may occur with Love test (extremely localized pain with the application of a pin or a ballpoint pen to the lesion and relief of pain on the removal of pressure), and the palpation of the site immediately next to the lesion does not cause any kind of pain. The patient had the classical triad of symptoms and a positive Love test. The contact of the lesion with ice worsens the pain. The Hildreths' test has 92% sensitivity and 91% specificity in the diagnosis of this lesion [9]. This test is positive if there is a reduction in pain and tenderness on exsanguination and ischemia of the affected limb.

Typically, these tumors have less than 1 cm in diameter and have a reddish or purplish color. They occur most frequently in the hand, especially in the nail bed. However, as about 25% are not subungual, their diagnosis is difficult. These lesions are benign and rarely malignant. However, when they are larger than 2 cm and histological features are suggestive of malignant tumor, metastases occur in up to 25% of cases [7]. Subungual glomus tumors occur more in females, but tumors at different locations have no gender predominance. These lesions may be single or multiple. The etiology of solitary tumors is not yet known; however, multiple lesions are associated with dominant autosomal mutations in the globulin gene located on chromosome 1p21-22 [10]. Most of these lesions occur in people aged between 30 and 60 years old and are extremely rare in childhood, perhaps due to the non-maturation of the glomus bodies and in elderly people due to atrophy and degeneration of these bodies.

Extra digital glomus tumors are difficult to diagnose, leading to long delays in diagnosis and misdiagnosis. The reasons are that they have unusual sites, and their clinical manifestations are usually different from those of classical glomus tumors. The incidence rates of pain and cold intolerance are lower in patients with extradigital tumors. Glomangiomas are the most common histological subtype of extra digital glomus tumors [11]. To avoid delay in diagnosis or misdiagnosis, when there are patients with painful or asymptomatic reddish or purplish subcutaneous nodules in extra digital location, glomus tumors should be included in their differential diagnosis.

There are diagnostic examinations that can support the diagnosis of this pathology. Ultrasound examination locates the lesion and determines the size and shape of the tumor. The Doppler ultrasound may be useful for detecting an increased blood flow. Magnetic resonance imaging is an excellent examination to characterize soft tissue lesions and is useful in the differential diagnosis of this pathology and other benign neural cell tumors such as schwannomas, neuromas, and neurofibromas. To the digital glomus tumors, MRI has a sensitivity of 90%, a specificity of 50%, a positive predictive value of 97%, and a negative predictive value of 20% [12]. This demonstrates that a negative image does not rule out a glomus tumor. MRI findings in extradigital glomus tumors are limited to case reports. However, the results seem to be similar [13]. These tumors must also be differentiated from other types, such as neuromas, the latter of which have also a positive Tinel test.

The treatment of choice for single lesions is surgical excision [14]. This treatment usually results in the complete resolution of symptoms, leading to an immediate improvement in the quality of life. Up to one-third of surgically removed tumors relapse within 2-3 years post-surgical removal [11]. These relapses occur mainly by incomplete removal of the lesions.

Conclusion

The rapid diagnosis of glomus tumors is important to avoid treatment delays leading to chronic pain. Surgical excision is the treatment of choice, with some relapses related to incomplete removal. It is important to consider these lesions in the differential diagnosis of subcutaneous lesions, especially when classic triad is presented, with pain, cold sensitivity, and point tenderness.

What is new?

The glomus tumors are rare, and 75% occur in hands. Therefore, extradigital tumors are very rare and difficult to diagnosis. In this article, the authors present a clinical case of a patient with a long time pain in his knee with a glomus tumor that failed to be diagnosed. It is an important article to show that we must think this tumor in the differential diagnosis of these lesions.

List of Abbreviations

MRI Magnetic Resonance Imaging

Conflicts of interest

The authors declare that there is no conflict of interests regarding the publication of this case report.

Ethical approval

Ethical approval is not required at our institution for publishing an anonymous case report.

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Summary of the case

1	**Patient (gender, age)**	Male, 67-year old
2	**Final diagnosis**	Glomangioma
3	**Symptoms**	Pain, cold intolerance, and point tenderness
4	**Medications**	-
5	**Clinical procedure**	Surgical excision
6	**Specialty**	Orthopedics