Journal of Research in
Orthopedic Science
May 2024. Volume 11. Number 2

Case Report

Intravascular Glomus Tumor: An Extradigital Glomus Tumor of the Elbow

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Citation Najd Mazhar F, Mohammad Nabi S, Emtiazi N, Mahmoudi Nasab O. Intravascular Glomus Tumor: An Extradigital Glomus Tumor of the Elbow. Journal of Research in Orthopedic Science. 2024; 11(2):115-118. http://dx.doi.org/10.32598/JROSJ.11.2.1435.2

http://dx.doi.org/10.32598/JROSJ.11.2.1435.2

Article info:
Received: 02 Feb 2024
Revised: 18 Mar 2024
Accepted: 28 Mar 2024
Available Online: 01 May 2024

Keywords:
Glomus tumors, Glomangioma, Extradigital glomus tumor, Intravascular glomus tumor

ABSTRACT

Background: Glomus tumors (GTs) are infrequent, benign soft tissue neoplasms originating from the thermoregulatory apparatus of the glomus body. Predominantly, these tumors manifest in the hand. Although uncommon, the occurrence of GTs in extra digital locations and their intravenous presentation is a seldom observed phenomenon, posing a diagnostic challenge.

Case Presentation: In this case study, we document the presence of an intravenous GT in the elbow region, which manifested as chronic elbow pain with an atypical presentation characterized by the ineffectiveness of various treatments.

Conclusion: Following surgical intervention and excision of the tumor, the patient experienced complete resolution of symptoms. Hence, despite the rare incidence of this tumor, it warrants consideration in cases of persistent and refractory pain.

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Introduction

Glomus tumor (GT) is, in most instances, a benign tumor originating from the glomus bodies, which are arteriovenous anastomoses crucial for thermoregulation [1]. Glomus bodies manifest as neuromyoarterial structures within the reticular dermis, predominantly distributed in regions such as the digits, palms, and soles of the feet [2]. Although most cases of this tumor occur in these areas, occurrences in atypical locations devoid of glomus bodies have been documented, leading to misdiagnoses, delays in diagnosis, and ineffective treatments [2]. Additionally, intravenous GT (IVGT) is a rare form of extra digital GT (EDGT) with reported cases primarily located in the upper limb's forearm region [1-12].

Case Presentation

A 42-year-old male presented at the Hand Clinic with enduring discomfort in the region of his left elbow. The pain had been persisting for the past two years, intensifying over the last two months, particularly during elbow flexion. Despite undergoing treatments such as physiotherapy, splinting, and anti-inflammatory medications, all of which proved ineffective in alleviating the pain, the patient reported no recent trauma or underlying rheumatologic disease. During the physical examination, a tender subcutaneous mass was observed, situated medially to the biceps tendon and 2 cm distal to the anterior elbow crease. There was no history of cold intolerance. Routine laboratory tests, along with a plain x-ray and magnetic resonance imaging (MRI) of the elbow, revealed no abnormal findings. However, a sonographic examination identified a well-defined, tubular, hypoechoic mass, measuring 14×5 mm, in the subcutaneous fat of the cubital fossa, leading to a probable diagnosis of neuroma (Figure 1). The patient underwent tumor exploration under general anesthesia with tourniquet application, revealing an intravenous round mass, which was excised by ligating both ends of the vein. The excised specimen was subjected to histologic examination (Figures 2 and 3), which confirmed the diagnosis of a GT through both histologic and immunohistochemical analysis (Figures 4, 5 and 6). Subsequently, the patient remained symptom-free, with no recurrence of pain or mass during the 7-month follow-up period.

Discussion

The initial documentation of an intravascular GT was attributed to Beham, who identified a lesion located in the forearm [3]. The majority of documented cases have been located in the forearm, with only one previous case reported in the elbow [4]. It is noteworthy that vascular involvement in this type of tumor is not confined to veins, as there has been a documented case of radial artery involvement in the wrist [13].

Dealing with persistent and resistant pain in extremities poses a persistent challenge in the field of orthopedics, encompassing a range of pathologies. GTs typically manifest with a classic triad of symptoms, including pain, pinpoint tenderness, and hypersensitivity to cold, recognized in 63% to 100% of cases. However, extradigital GTs (EDGTs), arising from various anatomical locations, present with diverse symptoms, thereby creating a significant diagnostic challenge [2]. In the case under consideration, the absence of reported sensitivity to cold, combined with exacerbated pain during joint movements, particularly flexion due to its proximity to the elbow joint, was noted. Notably, a pivotal aspect in the clinical diagnosis of these tumors is the incongruity between local pain and tenderness relative to the small size of the cancer [13].

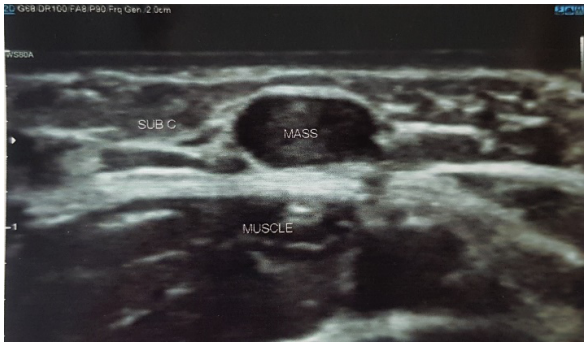

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Figure 1. The sonographic representation of the subcutaneous mass

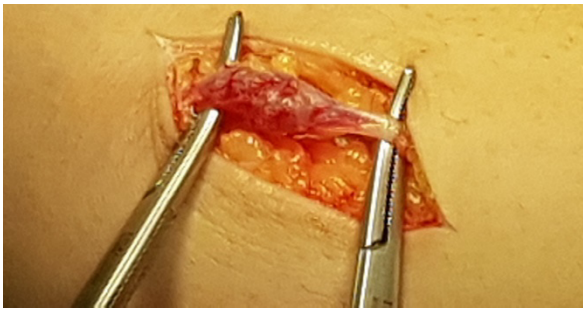

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Figure 2. The location of the tumor within the vein

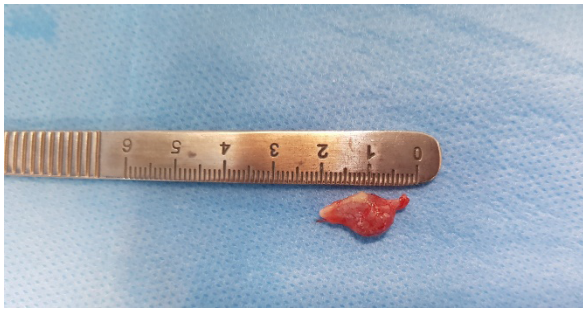

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Figure 3. The excised tumor encompassed by the vein

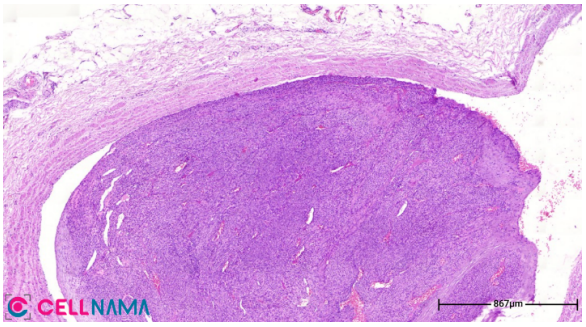

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Figure 4. A medium power view revealing a cohesive assembly of blue cells within a vascular space at ×20 magnification

The differential diagnosis of EDGT is contingent upon anatomical location, clinical symptoms, and the impact on adjacent structures. The diagnosis encompasses a spectrum of conditions, including hemangioma, vascular tumors, neuromas, neurofibromas, lipomas, paragangliomas, ganglion cysts, pigmented nevi, Pacinian corpuscle hyperplasia, and foreign bodies [2, 13].

MRI emerges as the most beneficial modality for diagnosing GT among various imaging techniques. The reported sensitivity and specificity of MRI for diagnosing GTs in the handstand are 80%-100% and 50%, respectively [9, 8, 13]. Previous reports on IVGT have highlighted the use of contrast-enhanced MRI in three studies, with diffuse homogeneous enhancement on postcontrast imaging being a common observation [5-7]. Furthermore, non-contrast enhanced MRI was utilized in one study and was reported as normal [8]. Before conducting an MRI scan, marking the tumor's location with a marker is advised, given the small size of the lesion. In instances where MRI was not employed, factors such as the superficiality and small size of the lesion led to its exclusion; this was mostly indicative of benign lesions, with surgeons opting for excisional biopsy for simultaneous diagnosis and treatment. Other diagnostic modalities, such as radiography, ultrasound, and angiography, have limited utility and are primarily used to rule out alternative diagnoses, occasionally causing confusion among surgeons [4, 8, 13]. A suggested straightforward diagnostic method involves the local injection of anesthetic around the area of maximum tenderness, resulting in complete symptom relief [4]. In patients without other medical issues, laboratory tests are generally close to normal and therefore offer negligible utility [2, 13].

Chronic pain and severe tenderness signify the most prevalent signs and symptoms of IVGT. Atypical presentations may lead to misdiagnosis and unnecessary treatment modalities, causing delays in final treatment that can endure for decades [7, 9]. Clinical examination and clinical suspicion continue to play pivotal roles in diagnosing these tumors. Contrast-enhanced MRI serves as a valuable diagnostic tool; ultimately, histopathological examination confirms the diagnosis. Recurrence is

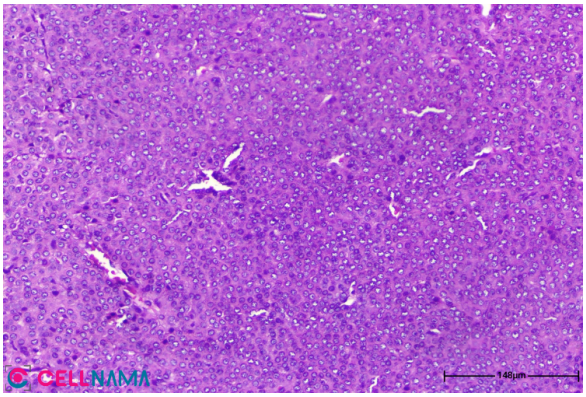

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Figure 5. A high-power view exhibiting clusters of glomus cells characterized by round, benign chromatin and minimal mitotic activity surrounding branching capillary-sized vessels at ×40 magnification

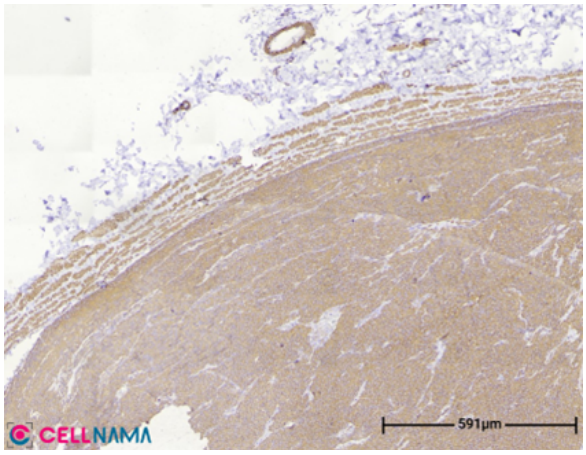

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Figure 6. A low-power view indicating SMA positive reactivity in glomus cells at ×20 magnification

infrequent and is typically attributable to incomplete excision. Moreover, should recurrence manifest, malignant tumors should be considered [8, 14].

The presentation of an exceptionally rare form of GT in an unconventional location underscores the significance of considering this tumor in the differential diagnosis for patients experiencing elbow pain. Familiarity with the relatively typical history and MRI findings offers substantial assistance in diagnosis and early curative treatment through surgical excision.

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this research. Written informed consent was obtained from the patient to publish this case report and accompanying images.

Funding

This research did not receive any grant from funding agencies in the public, commercial, or non-profit sectors.

Authors' contributions

All authors contributed equally to the conception and design of the study, data collection and analysis, interpretation of the results, and drafting of the manuscript. Each author approved the final version of the manuscript for submission.

Conflict of interest

The authors declared no conflict of interest.

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