

Atypically localized glomus tumors

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ABSTRACT

Aim: Glomus tumors are typically located in the subungual region in 75%-90% of patients. However, these tumors can be seen in atypical localizations which are extra-digital parts of the human body. Here, we present the management of a series of five patients with extra-digital glomus tumors treated surgically.

Patients and methods: The mean age of the patients was 40.6 years. The mean duration between symptom onset and presentation was 3.6 years. The localization of the tumors were anterolaterally of the thigh, posteriorly of the humerus lateral condyle, posteriorly of the humerus supracondylar region, anteromedially of the tuberositas tibia, and on the dorsal side of the wrist.

Results: The mean follow-up was at 52.8 months. For all patients, all lesions healed without any wound issues and no recurrences were noted during the follow-up period.

Conclusion: When a painful mass is found in the body, glomus tumors should be kept in mind. The consideration of symptoms, including pain, temperature sensitivity, point tenderness, and discoloration, common characteristics of glomus tumors, may aid diagnosis.

Key words: *Glomus, glomus tumor, atypical, extra-digital glomus tumor*

Introduction

Glomus tumors are rare benign tumors derived from the neuromyoarterial canal system of the glomus body that regulates the circulation of capillaries in the skin [1]. These tumors are typically located in the subungual region in 75%-90% of patients [2]. However, in the English literature, there are case reports and articles indicating that these tumors are seen in atypical localizations that are extra-digital parts of the human body [3-5].

Here, the management of a series of 5 patients with extra-digital glomus tumors treated surgically is presented. The importance of these patients is that the

diagnosis was delayed or even misdiagnosed. A review of the literature is also put forth.

Cases and Methods

A retrospective chart review of 5 patients with extra-digital glomus tumors over a period from June 2007 to August 2013 was performed. There were two female and three male patients with a mean age of 40.6 (range: 12-53) years. The demographic data of the patients are summarized in Table 1. The localization of the tumors were anterolateral of the thigh, posterior of the humerus lateral condyle (Figures 1,2), posterior of the humerus supracondylar region, anteromedial of

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Table 1. The demographic data, physical examination findings, and preoperative and postoperative VAS scores.

	Case 1	Case 2	Case 3	Case 4	Case 5
Gender	Male	Male	Female	Male	Female
Age (years)	47	41	53	12	50
Localization	Elbow (Posterior of the supracondylar region)	Elbow (Posterior of the lateral condyle)	Thigh (Anterolateral of the thigh)	Knee (Anteromedial of the tuberositas tibia)	Wrist (Dorsal side)
Mean duration between symptoms onset and presentation	5 years	2 years	8 years	1 year	2 years
Pain	(+)	(+)	(+)	(+)	(+)
Temperature sensitivity	(+)	(-)	(-)	(+)	(+)
Discoloration	(-)	(-)	(-)	(+)	(-)
Point tenderness	(+)	(+)	(+)	(+)	(+)
Preoperative VAS score	3	2	3	1	3
Postoperative VAS score	9	7	8	8	9

the tuberositas tibia, and the dorsal side of the wrist. Mean duration between symptom onset and presentation was 3.6 years (range: 1-8 years). All patients had paroxysmal pain and point tenderness (Table 1). Three of the five patients had cold sensitivity and one patient had discoloration of the skin (Table 1). There were no restrictions in range of motion (ROM) on the affected extremities.

The diagnosis of glomus tumor was based on a combination of clinical, radiologic, operative, and pathologic findings (Figures 1,2,3,4). Complete blood count, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), and electrolyte level analysis were performed in all patients. There were no significant results in the laboratory examinations of any of the patients.

Radiologic imaging comprising radiographs and magnetic resonance imaging (MRI) of the affected areas was conducted on all patients as part of the preoperative work-up. Direct radiographs did not reveal any outstanding features. MRI showed hypointensity in T1A-weighted sequences, hypersensitivity in T2A-weighted sequences, hypersensitivity in T2A FAT-SAT sequences, and also enhancement with contrast material (Figure 3).

After informed consent was obtained from all the patients, the mass was excised with an extended resection that included subcutaneous tissue.

Preoperative and postoperative pain assessment



Figure 1. A 41-year-old man with a mass on the posterior of the lateral condyle of the elbow.



Figure 2. Well-circumscribed glomus tumor is excised.

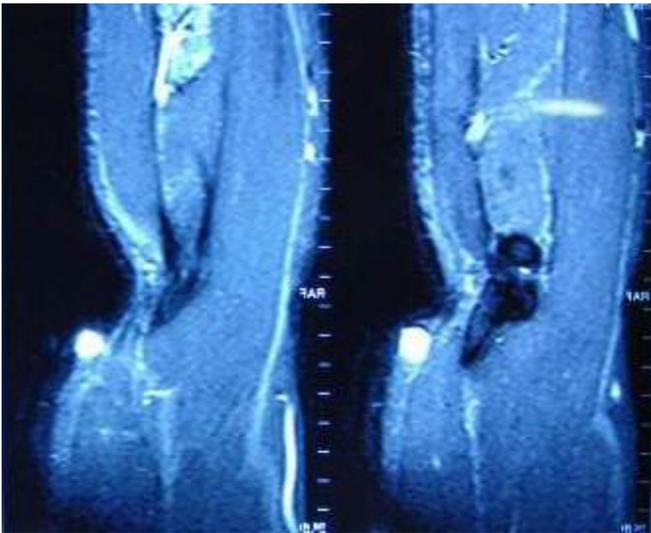


Figure 3. The MRI of the glomus tumor at the elbow.

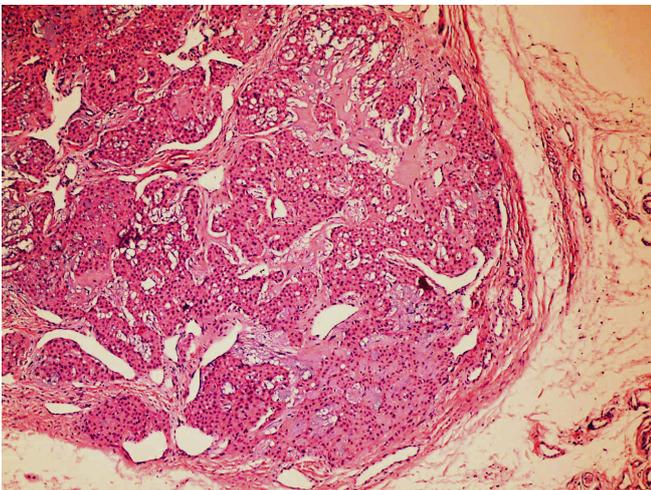


Figure 4. The microscopic image of a glomus tumor at the wrist of a 50-year-old woman.

comprised subjective evaluation with Visual Analog Scale (VAS) (Table 1). The preoperative mean VAS score was 2.4 (range: 1-3).

Results

The mean follow-up was 52.8 months (range: 18-92 months). All lesions healed without any wound problems and no recurrences were seen during the follow-up period. All patients were satisfied with the surgical outcome. All patients returned to work on the postoperative tenth day without need for job modification. The postoperative mean VAS score was 8.2 (range: 7-9).

Discussion

Glomus tumors, also known as solitary or solid glomus tumors, glomangioma, and nonchromaffin paraganglioma, were initially reported by Wood and in

1878, Kolaczek first described their typical subungual location [4]. The histologic diagnosis of glomus tumors was described by Masson [6]. Murray and Stout reported that the characteristic cell of the glomus tumor was the pericyte [7]. The pericyte was observed by Zimmerman and is found close to capillaries [8]. Pericytes are widely distributed and, according to Stout, this is why glomus tumors are able to arise in many tissues [9].

Although glomus tumors are generally seen in the subungual region, in the literature, these tumors are reported in the extremities, neurovascular system, and submucosal tissues [4,10-13]. Despite glomus tumors being believed to originate from Sucquet-Hoyer channels, the arterial segment of the glomus corpuscles and glomus bodies that control the blood flow in arteriovenous anastomoses, these tumors have been described in glomus corpuscles of free tissues, including the lungs, trachea, stomach, and fallopian tubes [5].

The histological assessment for the differential diagnosis of glomus tumors is essential and differential diagnosis of a painful nodule should include glomus tumor, vascular myoma, haemangioma, neuroma, angioleiomyoma, melanoma, and nodular hidradenoma [14,15].

Glomus tumors can be found singularly or in multiples, painful or painless, idiopathic or inherited autosomal dominantly [16]. Although digital glomus tumors usually more often occur in middle-aged women [1,17,18], extra-digital glomus cases are more frequent in men [13]. In the series here, three of the patients were male. The classical triad of glomus tumors is paroxysmal pain, temperature sensitivity, and point tenderness. Although the coexistence of these symptoms is often seen in digital glomus tumors, in extra-digital glomus tumors, these symptoms are rarely seen in one entity. According to a number of studies, the classical triad is observed in 63% to 100% of patients [5]. Except the classical triad, red or blue colored changes can be observed on the skin or nail depending on the depth of the lesion [19]. In this study, the coexistence of the classical triad symptoms was not identified. Yet, blue discoloration of the skin was seen in a 12-year-old boy with a subcutaneous localized glomus tumor at the an-

teromedial of the tuberositas tibia of the knee. As extra-digital glomus tumors are rare, it is difficult to diagnose. Van Greertruyden et al. stated that the diagnosis of these tumors may take up to ten years [1]. The mean time from onset of symptoms until diagnosis in the patients of this series was 3.6 years (range: 1-8).

Although several case reports have been reported thus far regarding extra-digital glomus tumors, the largest series was described by Schiefer et al. [5]. In that study, a total of 56 cases of glomus tumors were reported, with presentation by: three - hand, four - wrist, 11 - forearm, 4 - elbow, 4 - arm, 2 - shoulder, 1 - buttock, 5 - thigh, 10 - knee, 3 - leg, 2 - ankle, 2 - foot, 1 - back, 1 - nose, 1 - cheek, 1 - ear lobe, and 1 - trachea over the course of 20 years [5]. In the present series, a total of 5 cases of extra-digital glomus tumors were seen: 1 - wrist, 2 - elbow, 1 - thigh, and 1 - knee.

These tumors are rarely seen in the elbow [5,13,20,21]. The unique feature of Tomak et al.'s 56-year-old case is the location of the tumor was in the triceps tendon and the complaints of the patient began four years before diagnosis [21]. White and Jewer reported a 46-year-old male patient with a glomus tumor located at the anterior of the medial epicondyle and attempted to treat it based on a diagnosis of medial epicondylitis for a while [13]. The patients of this series had tumors localized subcutaneously, one at the posterior of the humerus lateral condyle and the other at the posterior of the humerus supracondylar region of the elbow. One of these patients had ongoing pain for 5 years and another for 2 years.

Glomus tumors had been described in the wrist within the literature [5,22-25]. Hoekzema et al. reported a 39-year-old interesting case. The primary feature was a large number of asymptomatic papular skin lesions had appeared on the patient's body during early childhood and, in subsequent years, on the left wrist and both thighs [23]. As a result of these skin lesions, glomus tumor was diagnosed [23]. The 50-year-old female in the current series had been misdiagnosed with tendinitis and treated with nonsteroidal anti-inflammatory drugs along with a wrist splint for 2 years because of the pain on the dorsal side of the wrist.

These tumors can also be seen in the thigh [5,23,26-

34]. Gonzales-Llanos et al. published a 50-year-old patient case report with a diagnosis of glomus tumor at the femur metaphysis [28]. In the literature, the most interesting glomus tumor of the thigh was seen in a 13-year-old girl diagnosed with calcified glomus tumor incidentally in the proximal femur, causing no complaints, after radiological examinations for idiopathic scoliosis [26]. Faggioli et al. examined a 24-year-old woman with a multifocal diffuse glomus tumor covering most of the right leg and, interestingly, this lesion was present since birth and had never manifested in any symptoms [27]. Similarly, the 53-year-old woman here was undiagnosed despite pain in the anterolateral of the thigh for 8 years at various hospitals.

The knee is the one of the most frequently reported regions of extra-digital glomus tumors [3,5,7-9,14,15,17,27,35-38]. The feature of Caughey and Highton's patient was non-diagnosis for 13 years. They reported hyperaesthesia of the skin over the patella but no other abnormality and radiographs had demonstrated minimal degeneration in the knee joint [8]. According to the authors here, the possible reason for the delayed diagnosis of Caughey and Highton's case was the available diagnostic tools, including MRI and ultrasonography (USG) were used in 1966. Although most of publications detailing glomus tumors of the knee are subcutaneous, intrapatellar or parapatellar localizations, Hardy et al. reported a case in the fat pad of the right knee of a 65-year-old man [35] and Kato et al. detailed a case with a glomus tumor beneath the plica synovialis of a 33-year-old man [36]. Bonner et al. reported an interesting case of a glomus tumor following total knee arthroplasty [14]. Although the patient had no pain at the postoperative first year examination, he did have a painful lesion at the anterolateral aspect of the knee joint diagnosed by ultrasound examination [14]. Panagiotopoulos et al. published a 20-year undiagnosed case with a painful glomus tumor at the medial joint line of the knee [15]. Lekehal et al. excised a glomus tumor at the popliteal fossa with tibialis nerve compression and it recurred a year later without malign transformation [37]. The present series case included a 12-year-old boy with glomus tumor at the anteromedial of the tuberositas tibia. The duration between symp-

tom onset and presentation in this patient was 1 year.

Extra-digital glomus tumors can indeed be seen in children. As mentioned before, there is the case of the 12-year-old boy as part of this series, a very rare age for extra-digital glomus tumors [1,17,18,26,38]. Öztekin reported a 9-year-old child with glomus tumor at the popliteal region mimicking Baker's cyst [38]. As much as is known presently, this is the youngest case in the literature. Another notable feature of this series' juvenile case is, besides combination of all symptoms of the classical triad, discoloration was detected.

Ultimately, when a painful mass is found in the body, glomus tumors should be kept in mind. The consideration of symptoms, such as pain, temperature sensitivity, point tenderness, and discoloration, the characterization of glomus tumors, will aid diagnosis.

Conflict of interest statement

The authors have no conflicts of interest to declare.

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