Glomus Tumor of the Scrotum: A Case Report and Mini-Review

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Abstract
Glomus tumor in the genital area is extremely rare, with an extensive search in the medical literature revealing only 1 case arising in the scrotum. They can be easily mistaken, both clinically and radiologically, for skin neoplasms or primary testicular tumors involving the scrotum. This report presents a case of a 54-year-old man who presented with a painful right scrotal swelling. Ultrasound suggested the possibility of an epidermal inclusion cyst. The excised mass was diagnosed as a benign glomus tumor. To the best of our knowledge, this is the second case of glomus tumor of the scrotum described in the literature. This report expands the differential diagnoses of scrotal neoplasms. Furthermore, essential guidelines necessary to distinguish glomus tumor from other lesions in the scrotum are discussed.

Introduction
Glomus tumors are rare mesenchymal neoplasms. They originate from glomocytes, modified vascular smooth muscle cells present in the walls of specialized structures engaged in thermoregulation (glomus bodies) [1]. Most often occur in the extremities, typically in the subungual region of the fingers and extremities, and rarely involve the scrotum [2].

Glomus tumor is a rare benign form of neoplasms that originates from the arterioneural structure called the glomus body [3]. The normal glomus body is usually present in the stratum reticulare throughout the whole body [4]. Glomus tumors are typically found in subungual areas, e.g. most commonly nails, finger tips and extremities, but rarely arise in other different areas of the body, e.g. the gastric antrum, trachea and glans penis [5]. Only 1 case of glomus tumor involving the scrotal skin has been previously reported and our case report is considered the second case in English literature [6]. We report this case of a rare location of glomus tumor and a brief review of literature on this neoplasm in order to raise awareness of this exceptionally rare tumor location whilst highlighting the clinicopathological features and differential diagnoses.

Hoyer first described glomus tumors in 1877, and Masson completed its description histologically in 1924 [7]. These tumors are usually small in size, single, of unknown etiology, and multiple lesions are uncommon constituting < 10% of all cases. Glomus tumors are more common in adults, particularly females. Patients usually seek medical help early because of the symptoms – most commonly involving pain, and temperature hypersensitivity. The mass is usually difficult to be identified on
exam because of its size [8]. In a recent case of a glomus tumor of the wrist, research that employed cytogenetic and spectral karyotypic analyses showed a novel rearrangement involving chromosome bands 1p13 and 5q32 [9].

Case Report

A 54-year-old male presented to the Urology Outpatient Department of our institution with a right painful scrotal swelling which he had for the past year. He complained of severe pain on slight touch, and had no other medical illness or previous history of similar episodes. The patient had undergone a follow-up scrotal ultrasound (fig. 1) that revealed a small hypoechoic oval structure (4 × 2 cm) in the wall of the right scrotum, with peripheral vascularity noted on color Doppler. The radiology report suggested the possibility of an epidermal inclusion cyst.

On physical examination, the right scrotum showed a tender subcutaneous whitish swelling (0.5 cm in diameter) on the right lateral aspect of the patient’s right scrotum. There were no signs of inflammation. The patient underwent a right scrotal nodule excision on May 24, 2015. He has been followed-up in the clinic since then for the past year with no signs of tumor recurrence and has not experienced any similar symptoms.

The histological examination revealed a single firm, tan fragment (0.5 × 0.5 cm). No skin was identified, and the cut surfaces were tan in color and firm in consistency. Bisected, entirely embedded in one cassette. Immunohistochemical stains were performed for AE1/3, Vimentin, SMA and CD34. The histomorphology and immunohistochemical staining profile supported the diagnosis of a scrotal glomus tumor.

Pathologic Findings

The surgical specimen consisted of a single fragment with 0.5 × 0.5 cm well-defined firm, tan lesion. Microscopic examination, at low power, showed a well-circumscribed, but not encapsulated, neoplasm covered by a thin layer of fibrous tissue (fig. 2). Some areas presented
an organoid or epithelioid pattern with nests of neoplastic cells (fig. 3). In other areas, tumorous cells clustered around thin, branching, and dilated vessels (fig. 4). The neoplastic cells had a monotonous appearance, without well-defined cell borders, and with abundant, light, eosinophilic cytoplasm and round to oval bland nuclei without nucleoli. Atypia, mitoses, intravascular growth, and necrosis were absent. Immunohistochemically, the glomus cells did not express cytokeratin AE1/AE3 and CD34 (fig. 5). In contrast, the same cells strongly expressed smooth muscle actin (fig. 6).

Discussion

Glomus tumor in the genitals and particularly in the scrotal region is a very rare entity with very few cases reported worldwide. While it classically manifests as a triad of pain, temperature hypersensitivity and tenderness [4]; not all patients present with all three, with pain being the most common symptom. Histologically, glomus tumors are modified smooth muscle cells that control the thermoregulatory function of the dermal glomus bodies, and comprise an afferent arteriole, anastomotic...
vessel and a collecting venule [7]. Enzinger et al. [10] categorized glomus tumors histologically into 3 groups including glomus tumor proper, glomangioma, and glomangioniomyoma.

Diagnosis of these patients is quite challenging due to the small size of the tumor and the unavailability of simple cost effective diagnostic investigations. In some studies and reported cases, ultrasound and MRI have been used to aid in the diagnosis, but nothing has proven to be specific and sensitive enough. The curative treatment is surgical excision regardless of its location [11].

Generally, the published literature seems to suggest that glomus tumors arising in various locations of the body are not clinically aggressive and display either no or low recurrence [9, 12]. To date, there has been no evidence of recurrence or metastasis in 1 year since our patient had the surgery. However, recent reports suggest that whilst it is rare for such a tumor to express clinically aggressive behavior, nevertheless, unusual histological features, including an infiltrative growth pattern, mitotic activity, and nuclear pleomorphism have been reported in a number of patients [13], and when metastasis does occur, this disease is often fatal, e.g. in the lung [14]. In addition, atypical presentations include multiple glomus tumors [15].

This is the second case reported globally of a glomus tumor involving the rare ectopic scrotal skin, and the first case to be reported in the Middle East and Eastern Mediterranean region.

Consent

The patient provided written informed consent for the publication of this case report and the accompanying images.

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