Case Report

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Glomus Tumor of the Vulva: A Case Report and Review of Literature

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ABSTRACT

A glomus tumor is a rare benign soft tissue neoplasm that originates from the neuromuscular cells of the glomus apparatus, a specialized arteriovenous anastomosis involved in thermoregulation. Although primarily found in the deep dermis and subcutaneous tissues of the extremities, typically in the subungual region of the fingers, it is occasionally reported to be found in the gastrointestinal, respiratory, and genitourinary tracts. In terms of female reproductive tract, glomus tumors have been described to be seen in the external genitalia, vagina, cervix, and myometrium. These tumors are often associated with localized intense pain at the tumor site, and surgical excision usually provides a definitive cure with permanent relief from symptoms. Herein, we present the case of a 39-year-old woman with longstanding vulvar pain and severe introital dyspareunia, who was diagnosed with a clitoral glomus tumor. The clinicomorphologic and immunohistochemical characteristics are discussed, along with a review of the literature.

Keywords: Glomus tumor, vulva, dyspareunia

INTRODUCTION

A glomus tumor is a rare benign mesenchymal neoplasm of the skin that arises from the glomus body.[1-3] The glomus body is a contractile, richly innervated arteriovenous anastomosis in the dermis of neuromyoarterial origin, which regulates blood pressure and temperature by controlling peripheral blood flow.[4,5] While frequently located in the subcutaneous tissue of extremities, glomus tumors account for less than 2% of all soft tissue tumors. They are most commonly encountered in the subungual region of the digits, where glomus bodies are normally present.[6] Rarely, glomus tumors have been reported in the mediastinum, lungs, intestines, bone, mesentery, heart, lymph nodes, trachea, stomach, oral cavity, neural tissue,[1,2,6-10] and also seen in genitourinary tract, including the clitoris, vagina, cervix, ovary, and periurethral tissue.[6,11-15]

Glomus tumors are composed of round cells with surrounding vascular spaces of varying sizes, along with glomus cells, vascular structures, and smooth muscle cells in differing proportions.[16] They typically present as solitary, painful nodules, although in some cases-particularly those arising in childhood-they may appear as multiple lesions and are occasionally familial.[2,5] Paroxysmal pain radiating from the lesion is a characteristic symptom. The diagnostic triad includes localized pinpoint pain, exquisite tenderness even to light pressure, and exacerbation of symptoms upon exposure to temperature changes, particularly cold (cold hypersensitivity).[7]

Clinical examination, conventional radiography, ultrasonography, and magnetic resonance imaging (MRI) can be utilized to support the diagnosis.[17] Complete surgical excision typically results in permanent symptom relief and a definitive cure. While the majority of glomus tumors follow a benign course, rare cases of atypical or malignant behavior have been reported.[18-20]

In this report, we present the clinicopathologic features of a patient diagnosed with a vulvar glomus tumor, along with a review of the literature.

CASE REPORT

A 39-year-old woman (G3 P2 A1) admitted to our institution with longstanding vulvar pain, severe introital dyspareunia, and a tender vulvar mass. Gynecological examination revealed a solitary, well-circumscribed, palpable subcutaneous mass in the clitoral region. The mass measured 1 × 1 × 1.5 cm, was soft, tan-brown in color, slightly mobile, and exquisitely

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tender to light pressure. No relevant findings were noted in the family history or laboratory evaluation. The mass was completely excised with a margin of surrounding normal tissue under local anesthesia. The specimen consisted of a well-circumscribed nodule measuring 1 × 1 × 1.5 cm in size. The sections revealed a yellowish, fleshy mass. Histopathological examination indicated that the tumor was composed of numerous blood vessels of varying sizes, surrounded by collars of glomus cells (Figure 1a). The tumor cells were round, with punched-out round nuclei and amphophilic cytoplasm (Figure 1b). Pleomorphism, mitotic activity, and necrosis were not observed. Immunohistochemical analysis revealed diffuse, strong immunoreactivity for actin, whereas S-100, EMA, cytokeratin, and CD34 immunostaining were negative. The diagnosis was confirmed as a "glomus tumor". The postoperative course was uneventful, and excision of the tumor provided permanent relief of symptoms. Informed consent was obtained.

DISCUSSION

Glomus tumors, first described in 1924 by Masson, are special perivascular neoplasms believed to originate from modified smooth muscle cells in the walls of specialized arteriovenous shunts involved in thermoregulation. The exact incidence is unknown; however, it is estimated to be 1.6%.[6] Glomus tumors typically present as painful, small blue-red nodules located in the deep dermis or subcutis of the lower or upper extremities. There appears to be no sexual predisposition, except for subungual lesions, which occur more frequently in females.[2] They are most commonly encountered in individuals in their third to fifth decade of life. These tumors may appear as solitary or multiple lesions, with multiple lesions being more frequently reported in young populations associated with neurofibromatosis type 1. Some investigators suggest an autosomal dominant inheritance with incomplete penetration, and a gene for inherited glomus tumors has been identified on chromosome 1p21-22.[5]

Hemangiopericytoma, leiomyosarcoma with epithelioid change, rhabdomyosarcoma, Ewing's sarcoma, and nodular hidradenoma should be considered in the differential diagnosis. The diagnosis is made histologically. Glomus tumors are divided into three groups: glomus tumor proper (the most common variant), glomangioma, and glomangimyoma, based on the proportion of glomus cells, vascular structures, and smooth muscle cells. Immunohistochemically, glomus cells are positive for vimentin, smooth muscle actin, and neuron-specific enolase, while they are negative for cytokeratin and S100, and variably positive for CD34, caldesmon, and calponin.

Glomus tumors typically have a benign course, but at times they can be infiltrative. Recurrence is rare and can usually be managed through conservative reexcision. Malignant glomus tumors are extremely rare. Clinically, they may extend into surrounding tissue or recur but seldom metastasize.[2] Deep location, a size over 2 cm, atypical mitotic figures (five or more mitoses per 50 HPF), and moderate-to-high grade nuclear atypia should be considered as criteria for malignancy.

A few cases of glomus tumors involving the female genital tract have been reported in the literature. Clinical and pathological features of vulvar,[12,13] clitoral,[14,15,26] periurethral,[14,23] and vaginal[20-22,27] glomus tumors have been documented as isolated cases (Table 1). In these reported cases, pain was the most common symptom. Most of the cases occurred in individuals in their third to fifth decade of life. While glomus tumors are most commonly encountered as solid masses, they can also present as cystic nodules or skin thickening.[12-14,20,22,24] Atypia or mitosis was not commonly observed in the microscopic examination of the tumors, although Suharwardy et al.[25] reported a case with atypia and mitosis. All reported cases were treated with local excision of the mass, as in our case.

Other gynecological glomus tumors include those found in the uterus,[28] cervix,[29] and ovary,[15,30] with one case associated with a teratoma.[8] The difficulty in diagnosis arises from the

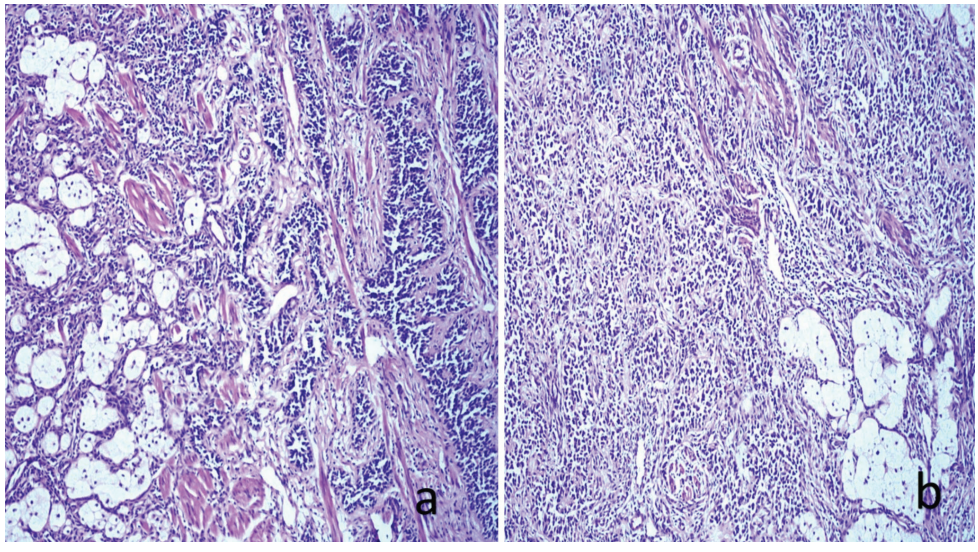

Figure 1. (a) Microscopic appearance of the tumor composed of numerous and varying sized blood vessels surrounded by collars of glomus cells. (b) Microscopic appearance of round cells of the glomus tumor with a punched out round nuclei and amphophilic cytoplasm

Table 1. Glomus tumor which reported by literatures

Study	Number of case	Age	Symptoms	Localization	Macroscopic pattern	Microscopic pattern	Diameter	Treatment
Banner and Winkelmann[21]	1	46	Vaginal tenderness, dyspareunia	Posterior vaginal wall (left)	Purplish-red, slightly elevated mass	Large vascularization, smooth muscle arranged in fascicles and about large vessels	1 cm	Local excision of mass
Katz et al.[12]	1	29	Intense pain	Labium minora (left)	Solid, extremely tender mass	Solid, angiomatous, tumor cells grew in cluster and associated with thin walled vascular spaces	1 cm	Local excision of mass
Kohorn et al.[13]	1	45	Pain, dyspareunia	Labium minus (left)	Redness area, slight thickening of skin	Solid No mitosis No pleomorphisim	0.5*0.5 cm	Local excision of mass
Sonobe et al.[14]	2	53	Dyspareunia	Clitoris	Oval, smooth outer surface	Solid,large vascularization The cells formed nests that were permeated by thin-walled masses network. No mitosis No pleomorphisim	2*1.5 cm	Local excision of mass
		46	Pain	Periurethral region	Solid	Solid No mitosis No pleomorphisim	3*2 cm	Local excision of mass
Moldavsky et al.[22]	1	45	Asymptomatic	Anterior vaginal wall	Solid angiomatous	Solid and angiomatous	1 cm	Local excision of mass
Malowany et al.[23]	1	61	Postmenopausal bleeding	Periurethral region	Focally ulcerated periurethral mass	Between the tumor cell nests and sheets were numerous capillaries and small blood vessels with associated edematous and hyalinized stroma. No areas of atypia, necrosis, or mitotic activity were seen.	1 cm	Local excision of mass
Mahapatra et al.[24]	1	39	Pain	Labium minus	Painful smooth swelling	The cells were arranged around the blood vessels at the periphery of the tumor. No atypia No mitosis	3*2*1 cm	Local excision of mass
Rahimi et al.[20]	2	53	Pain	Posterior vaginal wall	Solid mass	Small, circum- scribed clusters and nests or epithelioid cells were scattered throughout the neoplasm, between fascicles of smooth muscle cells No mitoisis, No necrosis	2 cm	Local excision of mass
		56	Asymptomatic	Anterior vaginal wall	Cystic nodule	tumor that contained small, circumscribed clusters and nests of bland epithelioid cells in a perivascular distribution	1 cm	Local excision of mass
Suharwardy et al.[25]	1	41	Painful mass	Clitoris	Firm, mobile, tender mass	Highyl vascular lesion, composed of plump cells, minimal atypia ,6 mitotic figures 50 high powered fields	1 cm	Excision of mass
Xie et al.[26]	1	36	Post-coital ache no pain	Glans clitoris	Tender mass, no ulceration	The vessels were separated by cells with mostly round uniform nuclei merging with spindle cells with fusiform nuclei and cytoplasmic eosinophilia	14*10*13 mm	Local excision of mass

unexpected locations of these tumors, as they can mimic nevi, cutaneous, adnexal, and neuroendocrine neoplasms.

Glomus tumors are most commonly found in the digits but can occur anywhere in the body. The combination of clinical examination and MRI enables early and accurate diagnosis of glomus tumors. Complete excision typically provides permanent relief of symptoms and an effective cure. Benign histopathologic features, including a uniform appearance with minimal atypia and a low mitotic rate, along with complete surgical removal, are associated with an excellent clinical outcome.

Ethics

Informed Consent: It was obtained.

Authorship Contributions

Surgical and Medical Practices: M.Ç.K., Concept: B.M., Design: B.M., Data Collection or Processing: M.Ç.K., Analysis or Interpretation: L.A., Literature Search: B.A., Writing: S.Ö.

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