Case Report

Glomus tumor in abdominopelvic cavity: a case report

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Abstract: To date, no report has been published about abdominopelvic glomus tumor, and the previous literature is lacking of Diffusion Weighted Imaging (DWI) and Perfusion Weighted Imaging (PWI) image interpretation. A 44-year-old female presented with mild abdominal distension recently due to a large irregular-shaped tumor in the abdominopelvic cavity during ultrasound examination. Abdominal computed tomography (CT) and magnetic resonance imaging (MRI) were performed, followed by a functional magnetic resonance scan. Consequently, the patient underwent complete resection of the tumor. No nodal metastases were found during imaging or surgery. The pathology report revealed a glomus tumor. We report the first female case of abdominopelvic glomus tumor supported by clinical imaging and pathological findings.

Keywords: Glomus tumor, abdominopelvic cavity, MRI, glomangioma, glomangiomyoma

Introduction

Glomus tumors are uncommon vascular neoplasms arising from vascular endothelial cells. Mostly, they are benign, but some cases with atypical/malignant behavior have been reported [1, 2]. Glomus tumors are divided into three types: proper glomus tumors, glomangioma and glomangiomyoma [1]. Classically, they occur under the fingernails, less commonly in the viscera, and are extremely rare in the abdominopelvic region. They occur in male and female with equal frequency, at between 20 and 40 years. Ten percent of the glomus tumors are multiple [2]. We report this case supported by representative radiologic findings, including CT, MRI and functional magnetic resonance reports, and correlate them with clinical pathology to facilitate early detection and differentiation of glomus tumors from other solid tumors.

Case report

A 44-year-old Chinese female farmer, with regular periods and 1 child, reported mild abdominal distension for a month. Medical ultrasound revealed a huge tumor in the abdominopelvic region, and the patient was admitted to our gynecological ward on February 15, 2016. Physical examination showed a huge palpable mass occupying the pelvic cavity, with the upper bound reaching 2 fingers above the umbilicus with a clear boundary. Ultrasound (GE, Voluson E6) investigations showed a huge, irregular and hybrid echo mass behind uterus containing several colored spots and slices represented by blood flow signals by color Doppler flow imaging (CDFI). CT (Siemens 64-slice, Perspective) revealed (Figure 1A, 1B) a demarcated and irregular soft-tissue mass in the pelvis and hypogastrium with uterus pushed to the lower left abdomen. MRI (Siemens, Skyra3.0T) revealed (Figure 1C, 1D) a cystic solid mass that was not distinct from the right margin of uterus. The solid portion was hypo-intense on T1WI, slightly hyper-intense on T2WI with obvious enhancement. No enhancement signal was found in the cystic area. Diffusion-weighted imaging (DWI) (Figure 1E) revealed moderate signal intensity in the solid portion. Perfusion-weighted imaging (PWI) (Figure 1F-H) showed rich but slow blood flow in the solid portion accompanied by a slowly arising intensity-time curve, which was similar to adjacent normal myometrium. The primary diagnosis was adnexal malignant tumor.

After general anesthesia, intraoperative exploration during abdominal surgery revealed a mixture of solid cystic mass measuring approxi-
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Figure 1. A plain computed tomography (CT) scan of the tumor revealing a clear and irregular contour, and a heterogeneous mass in the abdominopelvic cavity. The uterus is pushed to the lower left abdomen: (A) axial scan; and (B) coronal scan. A plain magnetic resonance imaging (MRI) scan reveals a cystic solid mass not distinct from the right margin of uterus. The solid portion was hypo-intense on T1WI (C), and slightly hyper-intense on T2WI (D). The signal is similar to the outer uterine muscle. (E) Diffusion-weighted imaging (DWI) shows moderate signal intensity of the solid portion (b = 1000). Perfusion-weighted imaging (PWI) shows rich and slow blood flow in the solid portion. Axial and T1-weighted images (F) with contrast demonstrate heterogeneous enhancement of the tumor. Regions of interest (G) on delayed phase contrast-enhanced axial MRI: the red ROI1 was represents the solid part of the tumor, while the green ROI2 denotes the myometrium. The gradual rise in the time-signal intensity curve (H) associated with the solid part of the tumor is similar to that of the adjacent normal myometrium tissue. (I) Microscopic examination shows numerous vascular spaces surrounded by uniform glomus cells (hematoxylin and eosin stain, × 200). (J) Immunohistochemical analysis of tumor cells reveals positively stained smooth muscle actin. (K) Immunohistochemical evaluation of blood vessels revealed CD34+ve staining.

mately 20 cm at its largest diameter. It was mostly well-circumscribed, without a clear boundary between the mass and the right broad ligament of uterus, as well as the isthmus uteri and part of mesentery. A radical resection was decided finally. The surgeon opened the tumor capsule from a portion of the mesentery, and sucked approximately 500 mL of transparent pale yellow liquid from the cystic portion of the mass. The shrinking mass was separated from the other tissue followed by complete hemostasis. Pathological analysis via optical microscopy and immunohistochemistry (Figure 1I-K) revealed a glomus
tumor characterized by abundant vessels surrounded by monotonous, small-to-medium-sized round cells with isomorphic round nuclei and eosinophilic cytoplasm. Immunohistochemical staining revealed the following results: desmin+, S-100+, ki67 1%, CD10+, SMA+, VIM+, CgA-, SYN-, inhibin-, CD34 (blood vessel)+, HMB45-, and AE1/3-. No nodal metastases were found during imaging or surgery. No recurrence was reported.

Discussion

Glomus tumors are an uncommon type of neoplasm occurring in the arteriovenous anastomosis. They consist of modified smooth muscle cells identical to those described in the glomus body [2], and constitute less than 2% of all soft tissue tumors [1]. Histologically, glomus tumor cells are arranged in sheets and nests around the expanding arteriovenous channels. The World Health Organization Classification of Tumors classified glomus tumors as hemangiopericytomas [3]. Histologically, glomus tumors were divided into three subtypes according to the proportion [1] of glomus cells, vascular and smooth muscle tissues into proper glomus tumors, which are the most common, glomangioma and glomangiomyoma. Clinically, glomus tumors typically occur in the subungual limbs of female patients [2], and rarely in the abdominopelvic cavity due to the lack of glomus bodies [4]. Two cases involving ovaries and one case associated with testis have been reported [5]. However, in these cases, the tumors were located in the gonads, without involving the pelvic cavity. In addition to gonads, the atypical locations include gastrointestinal tract [4], mediastinum [6], pulmonary organs [1], liver [7], trachea [8], and bone [9]. To our knowledge, this is the first report of glomus tumor associated with abdominopelvic region.

Compared with ultrasound and CT, MRI is associated with a superior advantage in delineating the lesion based on soft-tissue resolution. In this case, MRI showed a clear boundary demarcating the solid components of the irregular mass, which were closely attached to the uterus. The diagnosis is difficult, but still useful surgically. In addition to conventional MRI, functional MRI reveals greater details. Diffusion-weighted imaging (DWI) showed moderate signal intensity associated with solid portion, indicating closely packed tumor cells. Perfusion-weighted imaging (PWI) showed rich and slow blood flow in the solid portion with a slow rising, intensity-time curve similar to adjacent normal myometrium tissue. In this case, the rich vascular supply was similar to the glomus tumors in other atypical locations reported previously. However, the previous studies lacked data interpreted by DWI and PWI. Therefore, this is the first Diffusion-weighted imaging (DWI) and Perfusion-weighted imaging (PWI) imaging analysis of glomus tumors in atypical location.

Based on abundant vascular supply of glomus tumors, the differential diagnosis includes hemangioma, angiosarcoma, and hemangiendothelioma. Conventional imaging methodologies (including CT, MRI, and dynamic contrast-enhanced MRI) are useful in distinguishing glomus tumors from hemangioma or angiosarcoma. However, it is difficult to distinguish glomus tumor from hemangiendothelioma due to morphological similarities [7]. In this regard, immunohistochemistry plays an important role based on the staining features of hemangiendothelioma, which are double-positive for CD34 and vimentin, and negative for smooth muscle actin [7]. In contrast, glomus tumors are positive for smooth muscle actin [4-8]. In addition, a few neuroendocrine neoplasms should be regarded as differential diagnosis, since glomus tumors show neuro-endocrine function due to their unique anatomic location. The glomus tumors located in the thymus gland are similar to thymus carcinoid or ectopic pheochromocytoma in imaging manifestations [9]. They secrete catecholamines during via Indium-111 octreotide scintigraphy. Pathological HE staining suggested a diagnosis of glomangiomyoma in this patient.

Although glomus tumors are benign in most cases, the probability of malignancy exists [4]. The malignant transformation is roughly divided into two categories [10]: morphological similarity to leiomyosarcoma, and pleomorphic to -benign glomus tumor. It is mainly composed of round cells with highly malignant features microscopically, with frequent incidence of benign glomus tumors. No well-defined criteria are available to distinguish malignant glomus tumors from benign glomus tumors. A recent study [10] showed that a histopathology of malignant glomus tumor should be considered
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in any of the following conditions: (1) deep location (such as lung) and size > 2 cm; (2) atypical mitoses; or (3) marked nuclear atypia and 5 or more mitoses per 50 high-power fields. A further study with a larger sample size is needed to establish the standard of reference. As additional benign glomus tumors show inert growth pattern, their malignant potential warrants complete surgical resection as the basic treatment.

Therefore, lymphadenectomy is indicated for our case as needed, for instance, in the presence of intrapulmonary glomus tumor [1].

In conclusion, the patient presents with an insidious onset of glomus tumor located in the abdominopelvic cavity. Imaging examinations showed solid cystic mass with mostly clear border. The solid part was accompanied with rich and slow blood flow and mildly restricted diffusion. Glomus tumors carry the potential risk of malignant transformation. Atypical glomus tumors manifest varying morphological characteristics, along with a stable and abundant blood supply. Immunohistochemical analysis further supports the differential diagnosis. Complete surgical resection and regular follow-up are recommended as the basic treatment approaches for patients diagnosed with glomus tumor.

Disclosure of conflict of interest

None.

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