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Case Report

What´s the Harm in Postponing the Surgical Resection of Incidental Mesenteric Tumors? Report on the Management of a Rare Glomic Tumor in the Mesentery

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Abstract

Glomic tumors are vascular neoplasms arising from mesenchymal cells contained in glomic bodies. They affect mostly the acral soft tissue and rarely tissues with low concentration of these structures, such as those located in the abdominal cavity. This is the case report of a 78 year-old male patient, who presented with a long lasting abdominal pain, pointed out at the right iliac fossa, whose CT scan images showed a solid-cystic lesion in the mesentery. His surgery was scheduled to March, 2020. However, due to the onset of Sars-CoV-2 pandemic in Brazil, and the interruption by decree, of all non-emergency surgical procedures, including elective oncology ones, his surgery had to be postpone for half a year. He was finally operated in October, 2020. A laparoscopic surgical resection was successfully performed; the entire tumor was removed with no need for enterectomy. For our surprise, the pathological report, including immunohistochemistry staining, was compatible with a glomangioma from the mesenteric fat. No additional treatment was necessary, since it had no features for malignancy, and patient has been followed until today, with a recent CT scan (March, 2022) showing no signs of recurrence. Comparing this case with other scarce reports in literature, we conclude that no harm has been done, by postponing surgery. Although, this is not be followed as a general rule, considering that mesenteric tumors could be of uncertain malignant potential and only an accurate histopathologic analysis of the whole specimen can rule out malignancy.

Keywords: Glomangioma; Glomic Tumors; Glomus; Mesentery; Mesenchymal Neoplasm

Abbreviations

GIST: Gastrointestinal Stromal Tumor; GI: Gastrointestinal; ICU: Intensive Care Unit; HPF: High Power Field; IHC: Immunohistochemistry

Introduction

Mesenteric tumors are rare and include a diverse group of histological entities with different treatments and prognosis. Glomangiomas, also known as glomic (or glomus) tumors, are classically described in limb ends; their abdominal occurrence is exceptional [1]. The latters are rarely reported and consist of mesenchymal cells similar to modified smooth muscle cells of normal glomic bodies and glomic arteriovenous anastomosis, highly specialized in blood flow regulation for thermal homeostasis [2,3]. These tumors usually correspond to benign lesions, whose treatment basically consists of single surgical excision.

Although rare, the majority of intraabdominal glomus tumors occur in the stomach, and are phenotypically similar to the peripheral glomus tumors [4]. Other interesting and very unusual abdominal sites have been reported in recent years, such as in the kidney, liver, and peritoneum. In 2010 the first case of a large glomus tumor arising from the kidney was described, whose malignant potential was considered uncertain [5]. In 2011, the third case of a primary glomus tumor of the liver was reported in a female patient harboring a large abdominal cystic mass [6]. And in 2014, a peritoneal glomangiossarcoma was described in a 47-year old woman, following the investigation of a large bilateral parauterine mass, measuring 45 cm [7]. In all cases, the diagnosis could only be performed after surgery, based on immunohistochemistry, and the follow up showed no recurrence in nether of them. As far as we could find, reports of glomangiomas in the mesenteric have been published less than five times in medical literature over the last 50 years.

Case Report

A 78 year-old male, presenting with mild chronic pain in the right hemiabdomen, was initially investigated by computed tomography. As comorbidities, he was obese and hypertense, and had no significant family history of cancer. CT scan (Figure 1-A) showed a hypervascular mass of 60 x 48 x 40 mm, located in the mesenteric fat anterior and inferior to the right kidney. Among differential diagnosis, we considered a lesion of mesenchymal origin such as GIST, but also a carcinoid or desmoid tumor was suspected. Complementary endoscopic assessment was also performed. Colonoscopy was normal. In the upper GI tract, a 4mm polyp in the second portion of the duodenum was found. Endoscopic mucosectomy was performed at first, and the pathological analysis revealed a non-neoplastic polyp. Surgery was then indicated for the mesenteric tumor, and scheduled to late March 2020.

However, by that time, the city or Curitiba, southern Brazil, was struck by the first wave of the new Coronavirus (Sars-CoV-2) pandemic. For that reason, all elective surgeries had to be cancelled by decree, including oncological procedures, following a worldwide recommendation. In fact, the increase in the ICU and hospital admissions, due to respiratory failure was immense, in both private and public health systems. All resources were mainly destined to treat Covid-related patients and their complications.

Surgery was postponed for 7 months, until local authorities allowed back some categories of elective procedures, such as oncologic. In the meantime, patient remained home and managed to stay healthy. In October 2020, he underwent an uneventful laparoscopic resection, whose steps are shown in figure 2. During surgery we confirmed the position of the lesion, arising from the mesenteric fat, well vascularized by arterial branches. It was completely removed, extraction was protected by an adapted endobag (Figure 2-D). The specimen was reasonably soft (due to the cystic component) and surrounded by a capsule of connective tissue (Figure 3). Patient was discharged in the first post-operative day.

A careful pathological study was necessary, since hematoxylin-eosin staining (Figure 4) raised the suspicious of a low differentiated neoplastic lesion. It was composed of epithelioid cells with discrete atypia and pleomorphismus, a mitotic rate of 2/10 HPF, without atypical mitosis or necrosis, and no vascular or lymphatic invasion. The tumor was composed of a fibroid capsule infiltrating towards the adjacent adipose tissue, with free surgical margins. A large immunohistochemical panel was performed. It was negative for AE1/AE3 (carcinoma, epithelioid origin), S-100 (peripheral nerve sheath tumors), melan-A (melanocytic origin), RCC (clone PN-15, for renal cell carcinoma), and CD117 c-kit (GIST). The following markers were positive and are seen in figure 5: CD34 (positive in blood vessels), vimentin (positive in endothelial and mesenchymal cells), smooth muscle actin (focal positivity), Ki-

67 (2%), and calponin (focal positivity, spindle cells). This panel combination surprisingly confirmed the diagnosis of a glomic tumor (glomangioma), a neoplasia bearing a mesenchymal origin with solid epithelioid arrays associated with a strong vascular component.

No further treatment was necessary and patient is under surveillance ever since. He was recently evaluated in the outpatient clinic and new CT scan was performed, in order to assess the abdominal cavity. There are no signs of residual disease on the mesentery; image is displayed in figure 1-B. Despite that, patient still complains of eventual pain in the right iliac fossa, not related to any other anatomical findings.

Figure 1: A - CT scan prior to surgery, showing an heterogeneous irregular formation in the right hemiabdomen (arrow), diffusely enhanced by contrast in the portal phase, with lobulated and cystic inner formation, located in the mesenteric fat, apparently vascularized by branches of the superior mesenteric artery and drainage through the superior mesenteric vein; B - re-staging CT scan, 18 months after surgery, in which the mesenteric mass is no longer seen, but artefactual images related to laparoscopic clips (arrow).

Figure 2: Laparoscopic view of the mass in the mesenteric fat (A), following the steps of surgical resection (B), and final mesentery aspect (C), and the mass inside an adapted endobag (D) prior to extraction.

Figure 3: Entire surgical specimen (A) and the mass after longitudinal opening (B), where we can see the lobulated and cystic description found at CT scan.

Figure 4: Slide of hematoxylin-eosin stain at magnification of 100x, showing abundant blood vessels (larger structures filled with red blood cells), and diffuse proliferation of spindle cells among epithelioid cells widespread (pleomorphismus).

Figure 5: Immunohistochemical staining at magnification of 400x, showing strong vimentin marking (A), positivity of CD34 in blood vessels (B), focal positivity of smooth muscle actin (C), and focal positivity of calponin in spindle cells (D).

Discussion

Mesenteric neoplastic lesions are rare and usually accidentally diagnosed based on radiological findings, following investigation for vague abdominal symptoms. They include a diverse group of histologic entities. Most mesenteric masses are of lymphatic origin; the incidence of cystic mesenteric masses in the United States is estimated at 1/100,000, half of these are cystic lymphangiomas [1]. Treatment and prognosis vary according to diagnosis, being lymphoma the most common solid mesenteric tumor, followed by desmoid (whose annual incidence in general population is between 2.4 - 4.3 cases/100,000) [1]. Other types are exceptionally rare, based on sporadic reports.

A glomic tumor in the mesentery is even rarer, and may present clinically with a large abdominal volume, and pathologically with an infiltrative pattern, associated with necrosis, nuclear atypia and mitotic activity [1,2]. We presented a case in which patient had a chronic abdominal discomfort in the right iliac fossa. His investigation showed a mass in the mesenteric fat, capable to be completely removed laparoscopically. Final pathological analysis described the tumor as a glomangioma, with very low malignant potential, based on some points, such as 2% Ki-67 immunohistochemical positivity.

Glomus tumors and glomangiomas can be malignant, although characteristics of malignancy are still not thoroughly understood, mainly because of its rarity[8]. A number of features for aggressive behavior are associated with unfavorable outcomes and/or uncertain malignant potential and may be pointed out as: the occurrence of relapse over time, large tumor sizes (> 2cm, when located in digits), deep location into the soft tissue, atypical mitotic figures, moderate-to-high nuclear grade, and high mitotic index (>5 per 50 HPF) [8].

Anthonius de Bruin., et al. reported in 2008 a glomus tumor in the mesentery with atypical features in a 74-year old woman [4]. Similarly to our patient, she presented with a long history of abdominal pain. In physical exam, a 15 to 20 cm mass was palpable in the lower abdomen, showing a hypervascular signal at CT scan, with a fluid collection attached to the right abdominal wall, including the umbilicus. There was no clear origin of the tumor and no evidence for metastatic disease. Cytological analysis of the aspirated fluid did not reveal evidence of malignancy. After the aspiration, a fistula occurred at the umbilical scar. A median laparotomy was performed, and a large mobile tumor was seen attached only to the mesentery of the appendix. The tumor, along with the cutaneous fistula, was successfully resected and patient recovered well. Pathologic analysis found no necrosis but a significant mitotic activity, up to 9 mitosis/ mm^2. Immunohistochemistry revealed widespread expression of smooth muscle actin and partially of actin. Stains for CD34, desmin, pan-cytokeratin, S100, CD117, calponin, and epithelial membrane antigen were negative. Differently from their patient, we found positivity of CD34 in blood vessels and focal positivity of calponin in spindle cells.

Another rare occurrence of a visceral glomic tumor was found in the kidney and reported by Gill and van Vilet, in 2010 [5]. Their case also presented with a large mass; pathologically they found an increased mitotic activity and infiltrative margins, suggesting malignancy. Oppositely, in Iran, Geramizadeh., et al. reported a glomus tumor of the liver, to which they believed not to have evidence of malignant behavior. They described a 50-year old woman with a 15 cm hypervascular cystic mass in the left lobe of the liver, with fluid levels indicating intralesional bleeding, with multiple enhancing septa at CT scan. Differential diagnosis was made between hemangioma and hemangioendothelioma. After surgery, no atypia and no mitosis were identified. IHC of the tumor cells showed reactive smooth muscle actin, and CD34 was positive in both endothelial and myoid cells. Other markers, including S100, HMB45, AE1-AE3, CD10, Bcl2, EMA, CEA, chromogranin, NSE, desmin, CD99, and c-kit, were negative. The Ki-67 proliferation index was <1%.

Baleato-González., et al. [7], in Spain, performed laparotomy to excise a poor-defined abdominopelvic soft tissue mass with extensive areas of necrosis attached to the parietal and visceral peritoneum of the uterine fundus, terminal ileum loops and umbilicus, measuring 45 x 30 cm in diameter, with a smooth surface, shiny, very congestive and hemorrhagic. Microscopically, it had abundant capillary-sized vessels; at IHC, vimentin, (alpha)-smooth muscle actin, HHF-35 actin and collagen IV were stained intensely positively. Tumor cells were negative for desmin, CD31, CD34, cytokeratin AE1/AE3, melan-A and HMB 45. The final pathologic diagnosis was glomangiosarcoma. Despite the strong malignant potential, one year later, the patient remained asymptomatic without radiologic evidence of tumoral recurrence.

As we can see in all case reports, treatment of choice is surgery, whenever possible. Recurrences are not common and, if they occur, it indicates that either resection was inadequate or it was an infiltrating tumor from the beginning, in which positive microscopic surgical margins are likely to occur [3]. In cases of malignancy, delayed surgery will affect prognosis, due to local recurrence. The postponement of cancer surgeries in the last year affected all countries due to the overload on the health systems caused by the pandemic. Fortunately in this case there were no complications or disease progression during this period, and total resection was achieved with free surgical margins.

Conclusion

The pandemic caused by the new Coronavirus dissemination caused a delay in oncological diagnosis and treatment worldwide. Prioritization for elective surgeries had to be individualized. In this scenario, we emphasize the importance of ongoing elective surgeries and yet, the possibility of finding very rare oncological cases. Since glomic tumors are an unusual differential diagnosis of solid tumors of the abdominal soft tissues, most of the times benign, we experienced no harm by postponing surgery for this patient, in the context of the recent COVID-19 outbreak. However, we are aware to be lucky not going through adverse pathologic features. Alike trauma surgeries, oncological procedures should be top priority in any operation room, despite pandemics, always keeping every sanitary measure to avoid viral dissemination.

Conflict of Interest

The authors declare not to have any conflict of interest.

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