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Case report

Extrauterine adenomyomas presenting in a 47 year old woman with a previous cesarean section

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1. Introduction

Adenomyomas are benign tumors that contain benign endometrial glands and endometrial stroma bordered by leiomyomatous smooth muscle. These tumors usually originate within the uterus. They differ from adenomyosis in that they are well circumscribed masses that have discrete borders which separate them from surrounding tissue. Rarely, adenomyomas can occur outside of the uterus and even more rarely, outside of the pelvis. Pre-operative diagnosis of extrauterine adenomyomas based on imaging alone is very difficult, given that leiomyomatosis peritoneii and carcinomatosis can have similar radiographic features. It is only with histologic examination of the surgical resection specimens that pathologists are able to differentiate these tumors. In this report, we describe a case of extrauterine adenomyomas arising both inside and outside the pelvis in a 47-year-old woman with a history of a cesarean section. To the best of our knowledge this is the first case of extrauterine adenomyomas in a patient only with a history of a cesarean section and no other reproductive surgeries.

2. Case report

A 47-year-old woman, G5P1041 with regular menstrual cycles, presented to her benign gynecologist with hip pain. She was otherwise healthy with no medical problems. Her past medical history is only significant for a cesarean section more than 13 years ago. Pelvic exam revealed an 18 week, anteverted uterus with a palpable 5 cm left adnexal mass. The remainder of the physical exam was unremarkable. Magnetic Resonance Imaging (MRI) of the abdomen and pelvis showed an enlarged uterus with multiple myomas, a 3 cm complex mass of the right ovary, a 7 cm complex solid mass anterior to the rectum, and a left adnexal mass separate from the ovary(Fig. 1a and b). No lymphadenopathy was noted. Upon initial workup, her blood tumor marker CA 125 was 88.8 Units/mL (normal < 38 Units/mL). The patient was then sent to Gynecologic Oncology due to concern for leiomyomatosis peritoneii or carcinomatosis and subsequently underwent exploratory laparotomy during which total abdominal hysterectomy, left salpingectomy (right tube was absent), right ovarian cystectomy, and excision of multiple masses at the bilateral uterosacral areas and sigmoid mesentery were performed. Given the patient premenopausal state and grossly normal ovaries, oophorectomy was not performed. On gross examination, multiple tan, firm, whorled nodules were identified in the uterine myometrium without hemorrhage or necrosis, consistent with myometrial leiomyomas. One uterine mass (7.4 cm in size) and multiple extrauterine masses (ranging from 3.0 cm to 7.0 cm in size) located in the bilateral uterosacral areas and the sigmoid mesentery were sectioned to reveal a white-tan firm whorled cut surface with some soft areas and cystic changes filled with viscous yellow fluid. Histologic examination of the uterine mass and multiple extrauterine masses all showed benign endometrial glands and endometrial stroma bordered by leiomyomatous smooth muscle without cytologic atypia or necrosis, consistent with adenomyomas (Fig. 2a and b). No evidence of adenomyosis within the uterus was identified. The right complex ovarian cystic mass was a serous cystadenoma. The patient's postoperative period was uneventful and she was discharge on the first postoperative day.

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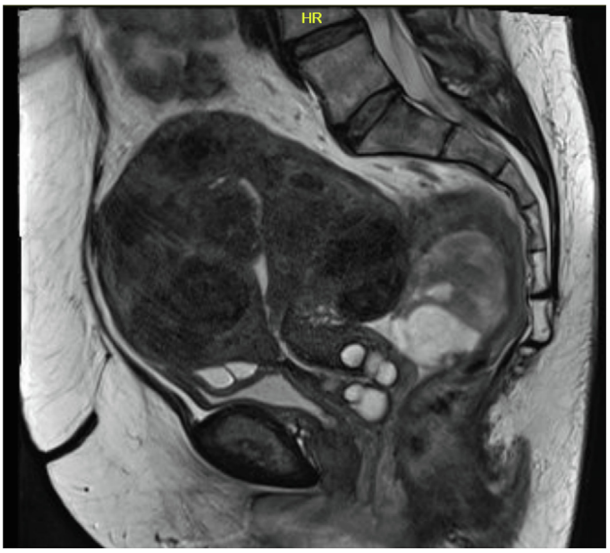

Fig. 1. a: MRI: T2 Sagittal Pelvis. b: MRI T2 Axial Pelvis.

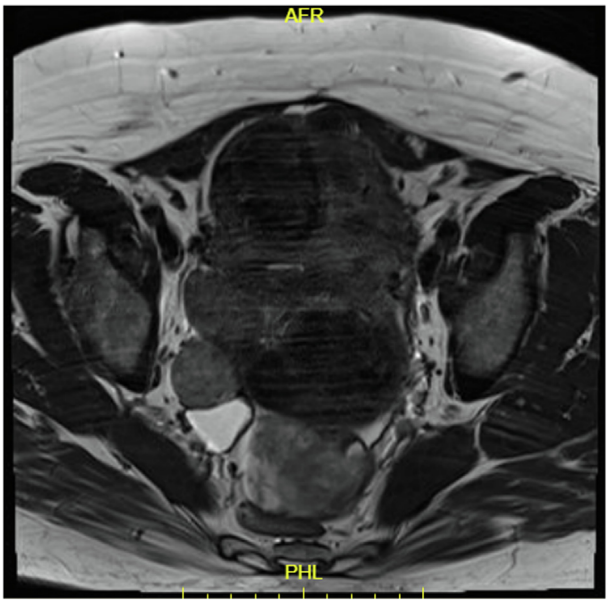

Fig. 1. (*continued*)

3. Discussion

To date, approximately 35 cases of extrauterine adenomyomas have been published in the literature based upon the most comprehensive review article to date (Paul et al., 2018) and PubMed search of articles published in the time since the review article publication (Belmarez et al., 2019). We present a case to further add to the knowledge of this rare finding in a patient with history only significant for a cesarean section.

The most common clinical presentation of extrauterine adenomyoma is abdominopelvic pain. While imaging is a typical part of a pelvic pain workup, as with our patient, there is currently no way to definitively diagnose benign adenomyomas with imaging alone. Due to this fact, patients are commonly sent to gynecologic oncology for care as the differential diagnoses of multiple extrauterine masses include pedunculated and/or necrotic fibroids, leiomyomatosis peritoneii and carcinomatosis as well as extrauterine adenomyomas. It is therefore important that gynecologic oncologists be aware of this rare possibility and understand the current theories of the pathophysiology behind it

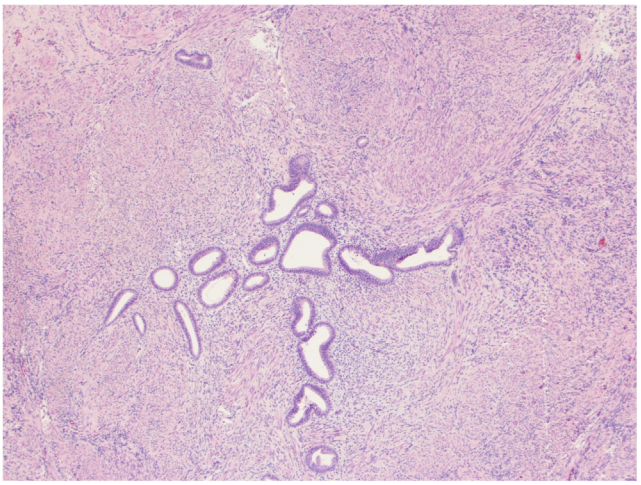

Fig. 2. a: Circumscribed sigmoid mesenteric mass composed of benign endometrial glands and endometrial stroma surrounded by abundant smooth muscle which is compatible with adenomyoma (hematoxylin and eosin stain; original magnification: 4×). b: Endometrial glands, endometrial stroma with surrounding smooth muscle cells (hematoxylin and eosin stain; original magnification: 20×).

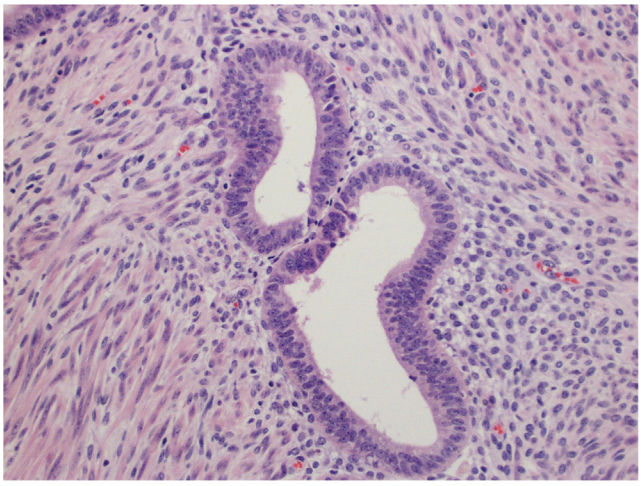

Fig. 2. (*continued*)

given the lack of current treatment recommendations when discovered. Surveillance and treatment to date have ranged from none to ovarian suppression with a GnRH agonist (similar to treatments proposed for another rare entity, disseminated peritoneal leiomyomatosis, depending on the presentation and which mechanistic theory is considered most likely, (Paul et al., 2018; Belmarez et al., 2019; Carinelli et al., 2009). There is currently no consensus by any medical group that we know of.

Currently there are five proposed theories for the pathophysiology of extrauterine adenomyomas. Cozzuto et al. published the first known case of an extrauterine adenomyoma in 1981. In this publication, he proposed the theory which focused on patients with a history of endometriosis (Cozzutto, 1981). It was proposed that adenomyomas form when there is a focus of endometriosis which undergoes metaplasia in to smooth muscle. While this held in his case report, future cases have reported extrauterine adenomyoma in patients without a history or evidence of endometriosis during surgery. Another counterargument to this theory critics have discussed is that the majority of the adenomyomas described in the literature contain a majority of smooth muscle with only small amounts of endometrium, such as our patient.

The second theory, Defective Mullerian Duct Fusion, was proposed

in 1982 by Rosai et al. after Cozzutto's article was published (Paul et al., 2018; Cozzutto, 1981; Rosai, 1982). He proposed that extrauterine adenomyomas or "uterus-like masses" developed due to a partial or complete fusion defect. These defects could lead to uterine duplications or atresia. When this defect creates a unicornuate uterus with a rudimentary horn, said horn can detach and implant elsewhere leading to a uterus-like mass. This theory has been supported by a few cases which the patient had concurrent renal and/or lower genital tract congenital abnormalities (Redman et al., 2005; Paul et al., 2018). However, recently more case reports have been published with no evidence of these congenital abnormalities, including our patient discussed here.

The next published theory to be presented was the theory of subcoelomic mesenchymal metaplasia (Redman et al., 2005). The subcoelomic mesenchymal layer sits below the mesothelial layer of the peritoneum and contains multipotent cells which could differentiate under hormonal (estrogen) prompting. The strength of this theory is that it explains why some of the described tumors in case reports have responded to hormonal treatment such as oophorectomy or GnRH agonists (Carinelli et al., 2009; Redman et al., 2005). However, with limited data on the recurrence rates of these rare tumors, broad generalizations cannot be made as to whether recurrence did not occur due to treatment or due to surgical excision of the masses.

The most recently published theory, Mullerianosis, was purposed by Batt in a letter to the editor in 2010 (Batt, 2010). He theorized that heterotopic müllerian-like organoid tissue of embryonic origin could develop within other normal organs during organogenesis which would explain müllerian tissue with no obvious source of dispersion. He did note that this theory only held if three criteria were met: absence of pelvic endometriosis; no communication of adenomyoma with endocervix, endometrium, and endosalpinx; no prior surgery to the reproductive tract. As such, our patient does not fit this theory either, as she previously had a cesarean section.

A theory which has not been formally published but has been commonly discussed regarding leiomyomatosis peritonealis is that of pelvic seeding during surgeries to the reproductive tract (Belmarez et al., 2019). It has been considered that during hysterectomy or myomectomy, cells can be seeded within the abdomen and pelvis which can lead to extrauterine leiomyomas, especially when morcellation without a bag occurs. By extension, it is feasible to extrapolate that extrauterine adenomyomas could occur by the same mechanism. However, in patients where both leiomyomatosis peritonealis and adenomyomas were found, the patients had no history of adenomyosis on pathology.

More cases will need to be reviewed to determine which, if any of the proposed theories is correct as none of the above theories described has a strong base given the relative paucity of information on this uncommon phenomenon. We proposed to add this case to the literature given that the patient has a common presentation on a rare event. Her case is interesting in that she has no history of endometriosis or müllerian abnormalities and had a cesarean section as her only abdominal surgery. To date the only surgical history mentioned in case reports as pertinent has been hysterectomy and myomectomy. In order for a clearer determination to be made on the etiology of this rare tumor more cases need to be reviewed to determine similarities and differences between patients. Once the etiology of these tumors is determined treatment options will follow. Currently no recommendations are available and there has been little follow-up on these patients and recurrence rates are unclear. As these patients commonly present to gynecology oncology services it is important for the gynecologic oncology community to remain up-to-date on the most commonly discussed theories so as to better discuss results and treatment options with patients who present with this rare finding.

4. Conclusion

In conclusion extrapelvic adenomyomas are exceedingly uncommon. We submit this case report to the literature in order to contribute to the overall fund of knowledge of this pathology which is as of yet of undetermined origin.

Author Contributions

W.M.B conceptualized the project, reviewed and edited. M.G wrote the original draft, reviewed and edited. X.W reviewed and edited.

Declaration of Competing Interest

Dr. Gruttadauria and Dr Wen have nothing to disclose. Dr. Burke reports other from Titan Medical, outside the submitted work.

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