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Case Report Open Access

Cellular Angiofibroma in the Space of Retzius: a Case Report

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Abstract

Background: Cellular Angiofibroma (CA) is an uncommon benign tumor that generally occurs in the vulvo-vaginal region in women. Extra-vulvar cases have previously been described but are exceptionally rare.
Case: A 45-year-old female with an incidentally noted pelvic mass in the Space of Retzius that was presumed to be a fibroid. Excision was complicated by significant haemorrhage from the surgical bed.
Conclusion: Extra-vulvar cases of CA are rare and have never been described in the Space of Retzius. We also discuss preoperative MRI findings to aid in diagnosis and surgical planning. Additionally, this CA was highly-vascularized, resulting in massive haemorrhage after removal. This is the first case to identify haemorrhage as a source of morbidity in CA.

Keywords: Cellular angiofibroma; Retropubic space; Benign pelvic mass

Introduction

Cellular Angiofibroma (CA) is a rare, benign, soft tissue neoplasm that was first described in 1997 [1]. It is a distinctive mesenchymal neoplasm comprised of spindle cells and prominent vasculature that presents almost exclusively in the vulvo-vaginal area in women. Subsequently demonstrated that CA occurs equally in men and is generally located in the inguino-scrotal region [2]. Extra-genital presentations of CA are quite rare and have been described in diverse locations including the pelvic retro peritoneum, chest wall and extremities [3-8]. In this case, we describe a patient with a large retro pubic cellular angiofibroma, whose excision was complicated by massive haemorrhage from the surgical bed.

This case was exempted from IRB approval at our institution.

Case

Clinical findings

The patient is a 45 y old nulligravid female with a history endometriosis and menorrhagia who presented for treatment of an incidental pelvic mass identified on Magnetic Resonance Imaging (MRI).

MRI revealed an enlarged, globular uterus with thickening of the junctional zone, consistent with adenomyosis. A heterogeneously T2 hyper-intense and avidly enhancing 6.6 × 5.5 × 6.2 cm mass was noted in the lower pelvis, inferior to the cervix and lateral to the vagina. It was presumed to be a subserosal cervical fibroid, although a definitive cervical or uterine attachment could not be delineated (Figures 1-3). Additionally, the MRI showed multiple T2 hypo intense masses consistent with small subserosal fibroids and a 10 × 12 cm cystic-appearing adnexal mass, most consistent with a cystadenoma.

Total abdominal hysterectomy, left salpingo-oophorectomy, right salpingectomy, extensive ureterolysis, retroperitoneal mass removal, and cystoscopy were performed *via* a 13 cm Maylard incision. Intraoperative findings demonstrated an anterior 6 cm solid mass in the space of Retzius displacing the cervix posteriorly. The cul-de-sac was completely obliterated by adhesions to the large ovarian cyst. There was significant peritoneal fibrosis, which was noted to overlie the ureters, requiring extensive ureterolysis to ensure appropriate visualization.

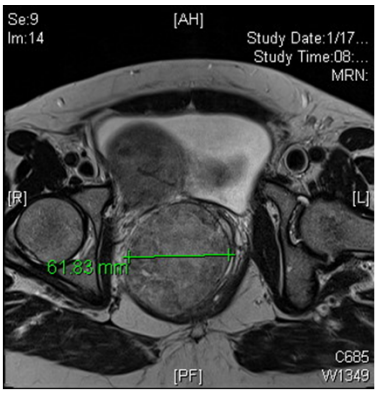

Figure 1: Axial T2-weighted MRI sequence of the inferior pelvis with retropubic mass

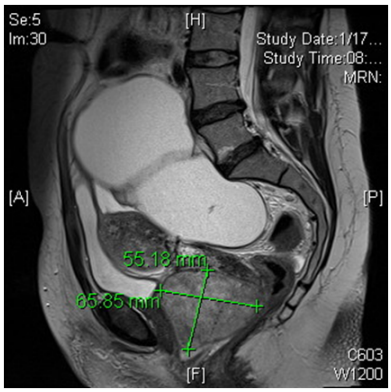

Figure 2: Sagittal T2-weighted MRI sequence of mass in inferior pelvis

Persistence of the 6 cm retropubic mass was confirmed following completion of the hysterectomy. The space of Retzius was then entered and dissected down bluntly to the level of the mass. The urethra was identified and noted to be deviated laterally. The mass was encapsulated and easily enucleated with blunt dissection. It was sent for frozen section analysis and was confirmed to be benign.

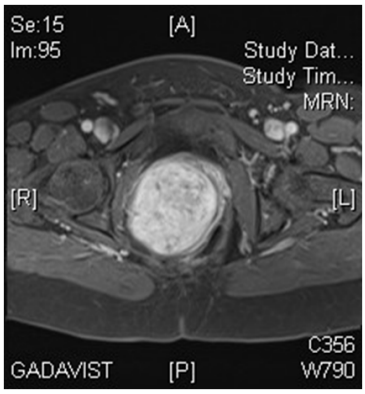

Figure 3: Axial fat-suppressed T1-weighted MRI sequence of mass in inferior pelvis with Gadolinium contrast enhancement

Brisk bleeding was observed immediately from the base of the mass. Given the location of the mass, urology was consulted intraoperatively. Hemostasis was ultimately achieved with suture ligation, as well as applications of Surgiflo™ (Ethicon, Cincinnati, OH), Tisseel™ (Baxter, Deerfield, IL), and Arista™ (Davol, Warwick, Rhode Island). Cystoscopy confirmed no evidence of bladder or ureteral injuries. After the case, the total estimated blood loss was 3.5 L. The patient received 6 U packed red blood cells, 1 U cryoprecipitate and 2 U fresh frozen plasma for intraoperative hemoglobin nadir of 3.7 g/dl, INR of 1.9 and fibrinogen of 53 mg/dl, respectively. Her postoperative course was uncomplicated and she was stable for discharge home by postoperative day 3.

Pathologic findings

The resected mass consisted of a 9.0 × 7.0 × 4.6 cm nodular portion of tissue with a smooth pink-tan external surface and a homogeneous, soft, tan-yellow, vaguely lobulated cut surface (Figure 4). Histologic sections demonstrated a low-grade mesenchymal neoplasm composed of bland spindled to stellate cells forming hypocellular edematous areas admixed with more cellular fibrous areas (Figures 5a and 5b). Numerous thin- and thick-walled vessels were regularly distributed throughout the tumor, some with hyalinized walls. No cytologic atypia or necrosis was identified, and mitotic figures were rare. A broad differential diagnosis of mesenchymal tumors was considered, including fibrothecoma, leiomyoma, endometrial stromal tumor, aggressive angiomyxoma and cellular angiofibroma.

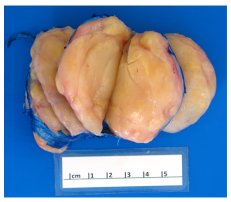

Figure 4: Space of Retzius mass after serial sectioning, demonstrating tan-yellow, vaguely lobulated cut surfaces with a soft consistency

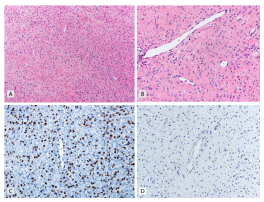

Figure 5: Space of Retzius mass, microscopic findings, photomicrograph courtesy of Dr. Borislav Alexiev. (a) Low-power hematoxylin-and-eosin (H&E)-stained section demonstrating uniform spindle cell proliferation with numerous interspersed vessels. (b) High-power H&E-stained section demonstrating bland spindled to stellate cells and variably sized vessels. (c) Immunohistochemical stain for ER demonstrating diffuse nuclear positivity in tumor cells. (d) Immunohistochemical stain for inhibin demonstrating complete lack of expression in tumor cells

A battery of immunohistochemical stains was performed to further characterize the tumor (Table 1). Positivity for Estrogen Receptor (ER) and Progesterone Receptor (PR) supported diagnosis of a gynecologic-type mesenchymal tumor (Figure 5c). Fibrothecoma was excluded based on negativity for sex cord-stromal markers (inhibin and calretinin), (Figure 5d). Negativity for smooth muscle markers (desmin, smooth muscle actin) helped exclude leiomyoma; this finding also favored a diagnosis of cellular angiofibroma over aggressive angiomyxoma. Positivity for WT-1 was also noted; though this finding is not typically associated with cellular angiofibroma, it has been described in the related entity of mammary myofibroblastoma [9]. Based on the overall clinic-pathologic findings, a diagnosis of cellular angiofibroma was rendered.

Antigen	Result in tumour cells
Vimentin	Diffusely Positive
ER/PR	Diffusely Positive
CD34	Negative
Inhibin	Negative
Calretinin	Negative
WT-1	Positive
S100	Negative
Pankeratin AE1/AE3	Negative
CD10	Negative
ALK-1	Negative
CD31	Negative
CD117	Negative

Table 1: Selected immunohistochemical results

Comment

Cellular angiofibroma remains a rare entity that classically presents in the vulvo-vaginal region in females and the inguino-scrotal region in males. While isolated extra-vulvar cases in females have been described arising in diverse locations including the retroperitoneum, lower extremities, chest wall and pelvis, this is an exceptionally uncommon presentation for this tumor [3-8].

Cellular angiofibromas of the vulva and perineum tend to be well-circumscribed with many demonstrating a fibrous encapsulation. Additionally, many are exophytic or pedunculated with a thin stalk that is amenable to surgical excision with a generous margin, which is the standard treatment for these lesions. Most of the previously described cases have evolved from superficial soft-tissues. In contrast, the origin of the mass described in this case was a deep pelvic space. This made removal and obtaining negative margins more technically difficult. While long-term follow-up of these lesions is limited, they appear to have very low risk of recurrence or metastasis when excised surgically [10], with only one prior case of recurrence described in the literature [6]. The spectrum of pathological characteristics is wider than initially appreciated [11], and cases with more cellular atypical as well as sarcomatous changes and elements have been described [7]. However, these changes have not been shown to confer additional risk for recurrence or progression.

There is only scant information published about imaging modalities in the diagnosis and management of CA, however MRI imaging played an important role in surgical planning in this case. On review of the literature, the only cases that have discussed MRI imaging in the management of CA have been in male patients. MRI plays an important role in delineating intra-and extra-testicular masses and can aid in surgical decision-making [12-15]. MRI findings in CA generally demonstrate T2 hyper intensity with a heterogeneous or in homogenous echo texture as well as intense contrast enhancement due to its rich vascularity [13]. Findings in T1 weighted modalities can be more variable because signal intensity is dependent on the fat content of the CA. They are generally T1 hypo intense with foci of increased avidity where fat content is denser [12]. The only other case in a female patient that utilized MRI imaging preoperatively had a similar preoperative radiologic impression of a fibroid or fibrothecoma [6]. While these tumours are much more common in the pelvic region than a CA, there are specific radiologic differences that can point to CA as a diagnosis.

Cellular angiofibromas are generally T2 hyper intense, whereas leiomyoma's rarely demonstrate this pattern. Fibroids can have some variability in MRI appearance but are classically T2 hypo intense, with areas of flow voids in the most hyper vascular masses. They are only hyper intense in cases of cystic degeneration [16,17]. Ovarian fibrothecomas can have an appearance similar to fibroids on MRI, but are distinct from the uterus. Cellular leiomyoma's, an uncommon subset of fibroids, can demonstrate a similar pattern to CA with T2 hyper intensity and contrast enhancement [17] and thus can be difficult to distinguish from CA on imaging alone.

Another unique aspect of this case was the presence of massive haemorrhage from the surgical bed at the time of excision. Despite the inherent prominent vascularity of cellular angiofibromas, no previously reported cases describe haemorrhage during mass removal. The anatomic origin of this mass may have contributed to the significant blood loss observed in this case. Bleeding in the retro pubic space particularly in the most inferior part of the pelvis can be difficult to manage given adjacency to the bladder and essential neurovascular structures. Most of the prior literature about bleeding in the retro pubic space comes from reports of complications of TVT, however isolated case reports describe haemorrhage associated with excision of other types of masses from this anatomic location [18]. The most important aspect is ensuring safety of other vital structures and postoperative cystoscopy to ensure integrity of the bladder and ureters.

In conclusion, extra-genital cellular angiofibromas remain a rare presentation of an already uncommon benign mesenchymal tumour. We describe a case of a CA in the retro pubic space, a previously undescribed location. MRI imaging can be helpful in differentiating these masses from fibroids and other pelvic masses, but given the radiographic similarities to cellular leiomyoma's, pathologic evaluation is essential to make a definitive diagnosis. Additionally, while bleeding complications have not been previously described in cases of CA, certain anatomic locations may predispose to haemorrhage at the time of surgical excision and should be considered as a possible source of morbidity in these patients.

Author Contributions

Nina Ayala: Wrote majority of case description and conclusion. Compiled additional sections from other authors.

Kerry Drury: Wrote introduction and participated in editing process.

Kruti Maniar: Participated in initial diagnosis. Wrote the pathology section and prepared images for manuscript.

Magdy Milad: Supervised manuscript production, and edited manuscript prior to publication.

Conflict of Interest

None of the authors have any financial disclosures or conflicts of interest to report.

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