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Cellular angiofibroma of the female pelvic cavity: a case report

Li Chen[1], Gumuyang Zhang[1*] and Hao Sun[1,2*]

Abstract

Background Cellular angiofibroma is a rare benign mesenchymal tumor that mostly occurs in the genital area. Its occurrence outside this region, particularly in the pelvis, is extremely rare. To our knowledge, this study reports the first case of cellular angiofibroma occurring in the pelvic cavity, except for one case reported in the retroperitoneum.

Case presentation A 25-year-old female patient with chronic, intermittent, dull pain in the lower abdomen that lasted for several months was referred to our clinic. Imaging studies revealed a tumor in the pelvic cavity anterior to the bladder. The radiographic characteristics of this tumor indicated a hypervascular nature, suggesting the possibility of a pheochromocytoma or a neuroendocrine tumor. The patient underwent surgical excision of the lesion. To date, no recurrence has been observed four months after excision.

Conclusions Cellular angiofibroma, although rare in the pelvic cavity, should be considered in the differential diagnosis of hypervascular pelvic space-occupying lesions. Immunohistochemical staining can help confirm the diagnosis of this condition. Treatment is generally straightforward, involving local excision of the tumor followed by postoperative monitoring.

Keywords Cellular Angiofibroma, Mesenchymal tumor, p16, Case report

Introduction

Cellular angiofibroma (CA) is a rare benign mesenchymal tumor that was first documented by Nucci et al. in 1997 [1]. It predominantly occurs in the genital area. CA commonly affects females in the vulva and vagina, while it affects males in the groin and scrotum. Although CA is rarely found in the pelvis, it has been reported in the retroperitoneum [2–4]. CA affects both genders, but it is more common in women, particularly those in their fifties [5]. The tumor manifests as a gradually growing mass and is often asymptomatic, although it may cause pain in some patients [6].

In this report, we present a rare and clinically atypical case of a 25-year-old woman with CA located anterior to the bladder in the pelvis. Additionally, we detailed the characteristics, imaging findings, histopathologic features, and treatment of the patient. To our knowledge, this is the second reported case of cellular angiofibroma occurring in a female pelvis.

Case report

This case report was approved for publication by the institutional review board. All of the patient details were anonymized to ensure patient confidentiality.

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A 25-year-old female patient with intermittent, dull pain in the lower abdomen for several months was referred to our outpatient clinic. She had regular menstrual cycles with no significant past medical or gynecological history.

The patient underwent computed tomography urography (CTU) in our clinic, along with a complete blood count, blood pressure and glucose monitoring, a 24-hour catecholamine assay, and biochemical examinations for tumor marker profiling. The results of these laboratory tests were within the normal range, and tumor markers, such as carbohydrate antigen (CA) 125, CA19-9, carcinoembryonic antigen (CEA), squamous cell carcinoma antigen, neuron-specific enolase, and lactate dehydrogenase (LDH)-199 were also within the established normal range. The CTU revealed an elliptical soft tissue density in the pelvic cavity, anterior to the bladder, measuring approximately 50 mm × 38 mm × 35 mm. The lesion had a relatively well-defined border with heterogeneous density on non-contrast imaging, characterized by multiple small cystic low-density areas. In addition, its solid component had a CT value of approximately 56 Hounsfield units (HU). During the arterial phase of the contrast-enhanced scanning, the lesion was significantly enhanced with a CT value of 149 HU, particularly within the small cystic areas. It was also significantly enhanced at the tail-like projections. In the portal venous and delayed phases, sustained enhancement with slightly reduced intensity was observed in the lesion, with multiple tortuous and dilated arteries and veins surrounding it. The CT values of the lesion were approximately 122 HU and 101 HU during the venous and delayed phases, respectively. This mass was found close to the anterior wall of the bladder, clearly demarcated from the bladder and adjacent bowel. The mass effect caused a posterior displacement of the anterior bladder wall (Figs. 1 and 2). Based on these findings, our institution's preliminary diagnosis was pheochromocytoma or neuroendocrine tumor. Consequently, the patient underwent robot-assisted laparoscopic pelvic tumor resection, and the tumor was completely removed.

Microscopic examination of the specimen showed a spindle cell tumor with dense cellularity, abundant eosinophilic cytoplasm, mild nuclear atypia, and a highly vascularized stroma with areas of hyaline degeneration. Immunohistochemical (IHC) staining revealed cytoplasmic positivity for CD34, smooth muscle actin (SMA), and β-catenin, while desmin, CD117, S-100, and DOG-1 were negative, with a Ki-67 index of 5% (Fig. 3). Based on the immunohistochemical results, the final diagnosis was confirmed as CA, and no tumor recurrence was observed during the four-month postoperative follow-up period, as confirmed by a repeat CT cross-sectional imaging.

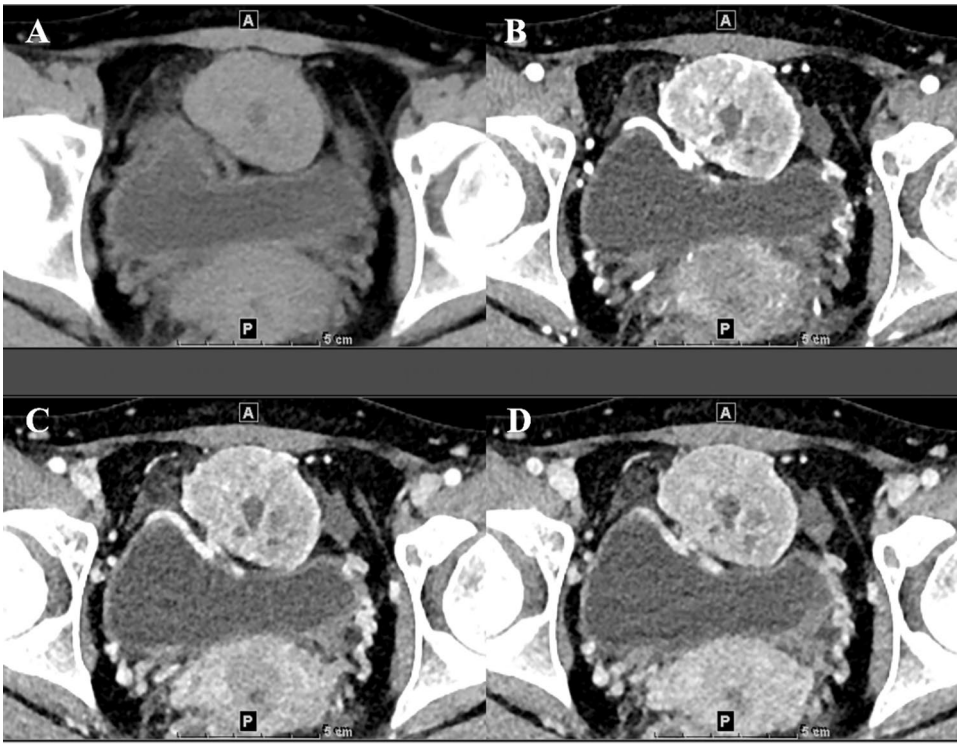

Fig. 1 The depicted images are as follows: (**A**) the non-contrast phase, (**B**) the arterial phase, (**C**) the portal venous phase, and (**D**) the delayed phase. The tumor exhibits a well-defined border with a heterogeneous composition. Its solid components were significantly enhanced during the contrast-enhanced scans, with a slight decrease in enhancement intensity over the phases

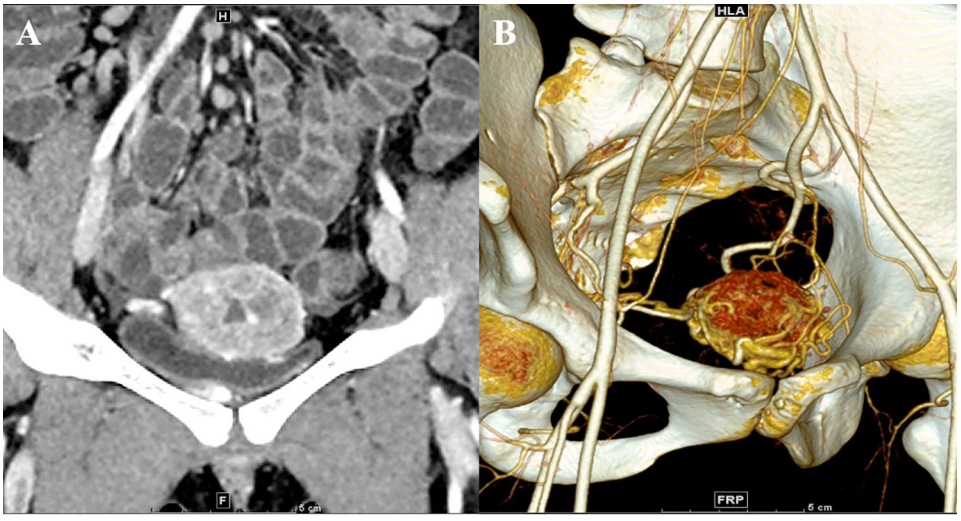

Fig. 2 (**A**) Coronal reformatted imaging of the tumor clearly delineated from the surrounding bowel and bladder, exerting significant pressure on them. (**B**) Volume rendering technique reconstruction reveals abundant vascularity within the tumor

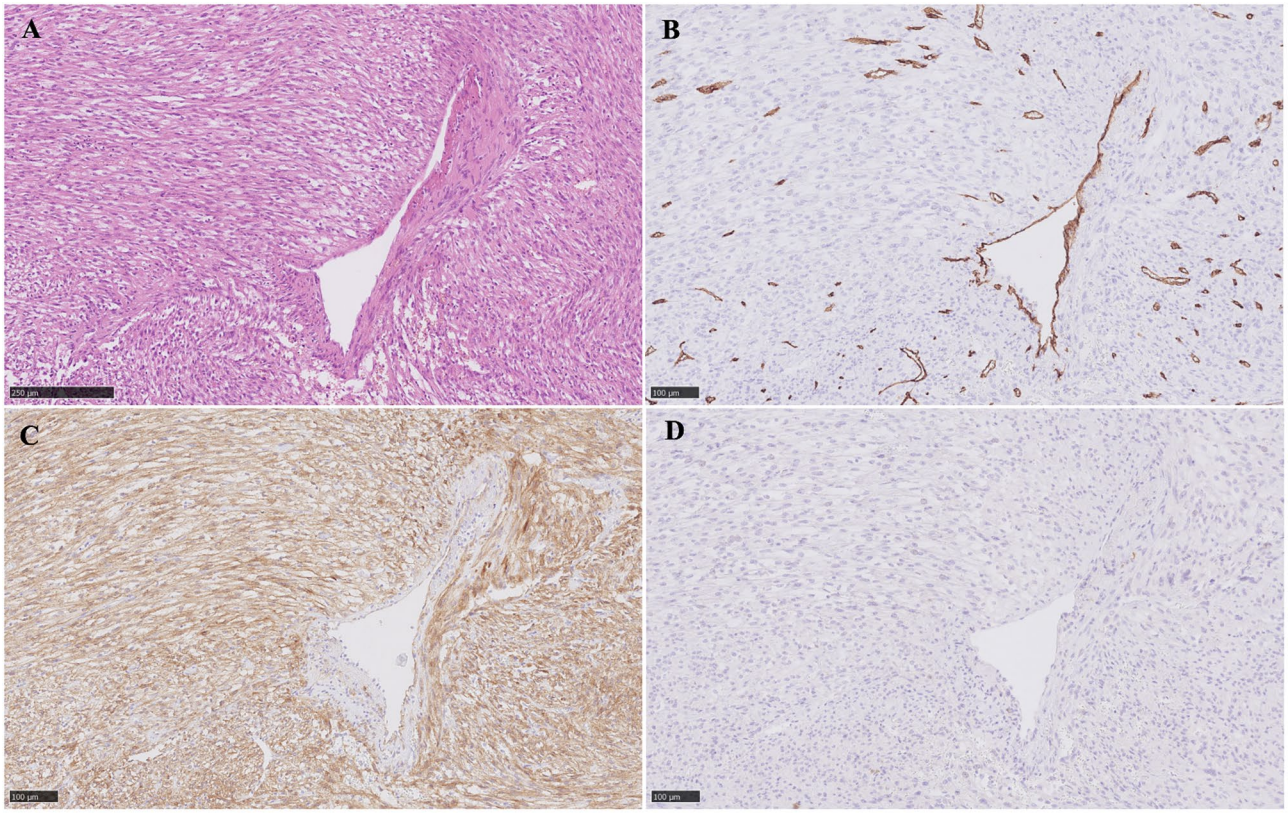

Fig. 3 **A** (magnification ×250): the hematoxylin-eosin staining specimen shows a spindle cell tumor with dense cellularity, abundant eosinophilic cytoplasm, mild nuclear atypia, and a highly vascularized stroma, with areas of hyaline degeneration. **B** (magnification ×100): immunohistochemical staining reveals cytoplasmic positivity for CD34. **C** (magnification ×100): IHC staining revealed cytoplasmic positivity for β-catenin. **D** (magnification ×100): IHC staining revealed negative for S-100

Discussion

Although studies have reported tumors occurring in the genital region, the etiology of CA is not fully understood [7]. Lane et al. reported a possible link between long-term estrogen therapy and CA development [8]. Some researchers have also proposed that CA arises from the differentiation of fibroblasts or myofibroblasts. Fluorescence in situ hybridization and other analytical

methods have been used to reveal deletions in the chromosomal region 13q14 in cases of CA, spindle cell lipomas, and mammary and vaginal myofibromatous tumors [9]. The similar histopathological features observed in this study suggest a potential link between these tumors [10].

Although CA mainly affects the vulvar, vaginal, and scrotal regions, rare cases have been reported in the oral mucosa, male pelvis, subcutaneous tissue of the chest wall, nasopharynx, retroperitoneum, and anorectal region [11–15]. CA is a well-circumscribed benign mesenchymal tumor, often located in the superficial soft tissues of the trunk [6]. Most cases of CA present as asymptomatic subcutaneous nodules, except for those in the nasopharynx and retroperitoneum. In the nasopharyngeal region, the tumor manifests as frequent epistaxis (nosebleeds) and persistent nasal congestion [12], whereas retroperitoneal tumors cause pain in the iliac fossa [2]. In this report, we presented the second known case of CA in a female pelvis with atypical clinical characteristics, manifesting only as mild lower abdominal pain.

The histopathological changes in CA include the presence of a well-circumscribed tumor located within the dermis and lacking a capsule [7]. In most patients, the neoplastic cellular component is predominant, with short, bluntly rounded spindle-shaped nuclei and sparse cytoplasm with indistinct cell borders. Additionally, medium-sized, thick-walled blood vessels with hyaline degeneration, slender collagen fiber bundles, and enlarged histiocytic cells are prevalent in this condition. Pseudo-vascular clefts may occasionally be observed, and mature adipocytes are often present. Similar to other tumors, focal cellular atypia was described in this report, and sarcomatous changes were also identified. IHC revealed that the tumor cells exhibited diffuse vimentin positivity, and CD34 was positive in most cases [7]. In female patients, some tumor cells showed partial positivity for estrogen receptor and progesterone receptor, while SMA, desmin, and S-100 were generally negative. However, in male patients, SMA, muscle-specific actin, and desmin may show partial positivity. The Ki-67 proliferation index is low.

CA often appears as a well-circumscribed, hypervascular tumor on CT or MRI images, with no other specific radiographic findings. It requires differentiation from the following conditions. ① Spindle cell lipoma: This tumor predominantly occurs in the posterior neck, shoulder, and upper back of males. The tumor comprises mature adipocytes and slender spindle cells in varying proportions, with myxoid degeneration in the stroma forming pseudo-vascular spaces. Spindle cells are CD34 positive, mature adipocytes are S-100 positive, and both are consistently negative for the Rb protein. Advancements in genetic testing are increasingly employing MDM2 detection to differentiate spindle cell lipoma, which typically lacks MDM2 gene expression, from malignant liposarcoma [16]. ② Angiomatoid fibrous histiocytoma: An uncommon soft tissue tumor of intermediate malignancy that commonly occurs in the external genitalia and perineal region, histologically characterized by multi-cellular and hypocellular areas, with prominent myxoid changes in the hypocellular areas. The tumor cells are epithelioid or spindle-shaped, clustered around blood vessels, and often multinucleated, with transparent or pink cytoplasm. They show strong positivity for desmin and vimentin. Angiomatoid fibrous histiocytoma is well-described to display a spectrum of variant histomorphologies, such as prominent myxoid stroma prompting, which often harbored a FET family gene EWSR1- CREM fusions [17]. ③ Retroperitoneal neurofibroma: It is located along the sympathetic ganglia in the paravertebral region. These tumors are well-circumscribed and variable in size, showing positivity for S-100. ④ Aggressive angiomyxoma: This tumor, often 10 cm or larger in diameter, exhibits infiltrative growth. It is composed of small spindle or stellate cells with lightly stained cytoplasm set within a myxoid matrix. These tumor cells express SMA and desmin. Additionally, based on accumulated case reports, Fluorescence In Situ Hybridization (FISH) analysis indicates that a chromosomal aberration at 12q15 results in the rearrangement of the high mobility group protein A2 (HMGA2) gene in nearly half of aggressive angiomyxoma cases [18]. Although its underlying mechanism remains unclear, it provides clues for distinguishing this tumor. ⑤ Labial Leiomyoma: This tumor consists of fascicles of spindle cells with well-defined cytoplasmic borders and cigar-shaped nuclei, which are positive for SMA [19].

Diagnosing CA based on clinical symptoms and imaging findings before surgery is challenging due to the absence of specific signs. Definitive diagnosis depends on needle biopsy or surgical excision followed by pathological examination. However, CT or MRI can indicate the extent of the tumor and guide the best surgical approach for complete lesion removal.

In summary, CA, although rare in the pelvic cavity, should be considered in the differential diagnosis of hypervascular pelvic space-occupying lesions. Most cases of CA exhibit benign behavior and are easily excised without showing atypia, and follow-up studies show low recurrence and no metastatic potential [20–22]. However, Hanae et al. suggested that previous research might have underestimated the recurrence or metastasic potential of CA, particularly in specific populations such as pregnant women, emphasizing the need for long-term follow-up studies [2]. The limitation in this case is the short follow-up period.

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Author contributions
L. C. and G. Z. contributed to writing original draft preparation. H. S. contributed to the idea design of the paper and the modification of the final draft. All authors reviewed the manuscript.

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Data availability
The data used during the current study are available from the corresponding author of reasonable request.

Declarations

Ethics approval and consent to participate
The Pecking Union College Hospital Institutional Review Board approval was obtained. Written informed consent was obtained from all participants before their inclusion in the study.

Consent for publication
Written informed consent for publication was obtained from the patient for their personal or clinical details along with any identifying images to be published in this study.

Competing interests
The authors declare no competing interests.

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