

Angiomatous leiomyoma of the urachus: A rare entity masquerading as extraluminal gastrointestinal stromal tumor

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Abstract

The urachus is a vestigial structure located between the dome of the bladder and the umbilicus, which results from the involution of the allantoic duct and the cloaca. Persistence of an embryonic urachal remnant can cause various problems during childhood and young adulthood. Urachal leiomyoma is a rare entity with very few cases being reported in literature. It can be misdiagnosed and confused with a wide spectrum of intra-abdominal or pelvic disorders. We hereby report a case of angiomatous leiomyoma originating from the urachal remnant in a 45-year-old lady, masquerading as extraluminal gastrointestinal stromal tumor. Understanding the embryological basis of these urachal disorders and their imaging features coupled with histopathological examination is crucial for the correct diagnosis and management. Pathological diagnosis is required to optimize the surgical approach and preclude unnecessary radical surgery.

Key Words: Urachus, leiomyoma, angiomatous

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INTRODUCTION

The urachus is a vestigial structure located between the dome of the bladder and the umbilicus which results from the involution of the allantoic duct and the cloaca.^[1] Persistence of an embryonic urachal remnant can be a source of infection and a nidus for the development of various benign and malignant neoplasms. Urachal leiomyoma is a rare entity with very few cases being reported in literature.^[2] It can be misdiagnosed and confused with a wide spectrum of intra-abdominal or pelvic disorders.^[3] We hereby report a case of angiomatous leiomyoma originating from the urachal remnant masquerading

as extraluminal gastrointestinal stromal tumor (GIST). To our knowledge, this is the first reported case of urachal angiomatous leiomyoma.

CASE REPORT

A 45-year-old lady came with a gradually increasing painful mass in the lower abdomen for the past six months with recent onset of increased frequency of micturition. There was no history of vomiting, weight loss or alteration in the bowel habits. She was a known case of diabetes mellitus well controlled on insulin, and had undergone tubal ligation 15 years back. There was no significant family history. Her general physical examination was unremarkable. On per abdominal examination, there was tenderness in the right iliac fossa with a mobile palpable mass measuring 5 × 3 cm in size. There was no regional lymphadenopathy. Her routine biochemical and hematological investigations were within normal limits, but she was found to be HBsAg positive. Computed tomography (CT) scan of the abdomen and pelvis revealed a well-defined heterogeneous, mildly enhancing, predominantly hypodense

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lesion measuring 5.8 × 3.6 cm lying anterosuperior to the bladder on the right side and in close proximity to the right rectus muscle [Figure 1]. It was thought to be an extraluminal GIST arising from the terminal ileum. The patient was undertaken for diagnostic laparotomy. A lower midline abdominal incision was given. The abdomen was opened in layers. A fibrous band was seen extending between the anterior dome of the urinary bladder and the anterior abdominal wall at the umbilicus, which in its central portion showed a 5 × 3 cm well-circumscribed lesion. It was thought to be an urachal remnant and was excised by ligating both its ends and was sent for histopathological examination.

The specimen consisted of a well-encapsulated nodular mass measuring 6 × 5 × 3.5 cm. The external surface was smooth with few congested blood vessels. The cut surface was solid homogeneous with gray-white appearance [Figure 2]. Microscopic examination revealed a well-circumscribed smooth muscle tumor comprising of spindle-shaped cells arranged in interlacing fascicles and whorls. The cells had long slender bipolar cytoplasmic processes and cigar-shaped nucleus. The stroma showed myxoid and hyaline change at places. Numerous thick-walled blood vessels were evident with their inner muscle layer arranged circumferentially and outer

layer blending with the less well-ordered smooth muscle of the tumor [Figure 3]. Immunohistochemically, the tumor cells were positive for smooth muscle actin (SMA) and desmin; and negative for CD117 (to differentiate from GIST). The vessels showed immunoreactivity for CD34 [Figure 4]. The patient is symptom free with no residual disease or recurrence after a follow-up period of 6 months.

DISCUSSION

The obliterated urachus, also known as median umbilical ligament, extends from anterior dome of the bladder towards the umbilicus, and measures 3-10 cm in length and 8-10 mm in diameter. It lies between the transverse fascia and parietal peritoneum in the space of Retzius.^[3] The urachus is a muscular tube with three distinct tissue layers: An epithelial canal with transitional (70%) or columnar (30%) epithelium; a submucosal connective tissue layer; and an outer layer of smooth muscle.^[4]

Anomalies related to urachal remnants most often occur in children, but they can also first present in adults. They include

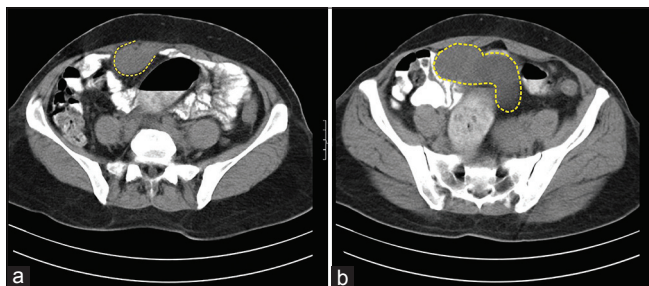


Figure 1: (a) CT scan of the abdomen and pelvis revealing a well-defined heterogeneous, mildly enhancing, predominantly hypodense lesion measuring 5.8 × 3.6 cm attached to the anterior abdominal wall at the umbilicus (yellow dotted line). (b) CT scan revealing the mass lying anterosuperior to the bladder on the right side and in close proximity to the right rectus muscle (yellow dotted line)

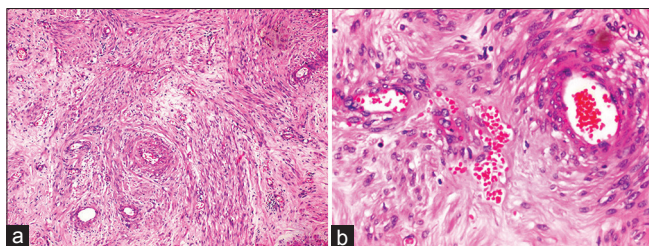


Figure 3: (a) Microscopic examination revealing a benign tumor comprised of spindle-shaped cells arranged in interlacing fascicles and whorls. The stroma shows myxoid change along with numerous thick-walled blood vessels (H and E, ×100). (b) Numerous thick-walled blood vessels are evident with their inner muscle layer arranged circumferentially and outer layer blending with the less well-ordered smooth muscle of the tumor (H and E, ×400)

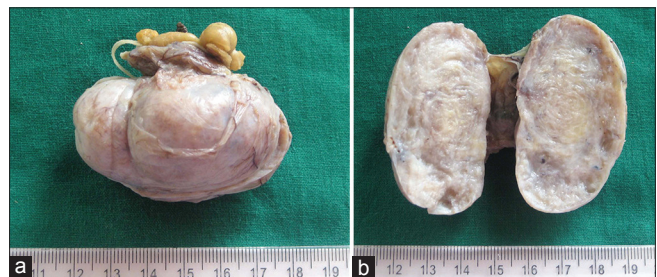


Figure 2: (a) External surface of the lesion showing a well-encapsulated smooth nodular mass measuring 6 × 5 × 3.5 cm, with a few congested blood vessels. (b) The cut surface of the mass showing solid homogeneous gray-white appearance

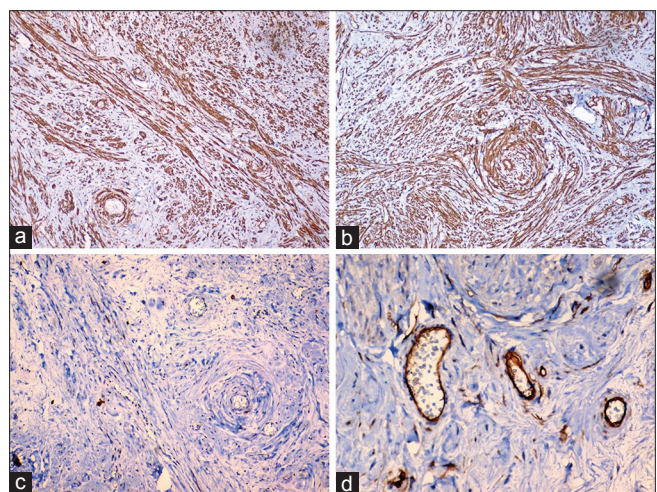


Figure 4: Immunohistochemically, the tumor cells show positivity for (a) SMA (×100), and (b) desmin (×100), and (c) negativity for CD117 (×400), (d) the vessels show immunoreactivity for CD34 (×100)

the following: patent urachus, blind sinuses in the anterior abdominal wall, urachal cysts and diverticulum, abscesses and granulomatous omphalitis.^[3,5] Benign urachal neoplasms including adenomas, fibromas, fibroadenomas, fibromyomas and hamartomas are extremely rare.^[4] Other benign tumors like desmoid tumor, inflammatory myofibroblastic tumor and teratoma arising from the urachus have also been reported.^[6,7] Leiomyoma of the urachus is a rare entity with only seven cases being reported.^[2] Its incidence is suspected to be higher, but this tumor type is detected only when complications such as bleeding or infection arise.^[8] Typical symptoms include lower abdominal pain, palpable mass and abnormal voiding.^[2] Ninety percent of the urachal tumors arise in the juxtavesical portion of urachus. The remaining 10% are located in the middle of the urachus or near the umbilical end. Some tumors deviate towards the right or left paramedian portion from midline; because during embryonic development, the urachus occasionally deviates from the midline to merge with one of the obliterated umbilical arteries.^[4] This can explain the right-sided location of the tumor in our case.

Leiomyoma arises from the outer layer of smooth muscle of the urachus and is comprised of spindle-shaped cells arranged in interlacing fascicles. Angiomyoma in addition shows the presence of thick-walled vessels with partially patent lumens. Typically, the inner layer of smooth muscle of the vessel are arranged in an orderly circumferential fashion, and the outer layers swirl away from the vessel, merging with the less well-ordered peripheral muscle fibers. Areas of myxoid change, hyalinization, calcification and fat can also be seen.^[9] The above findings were seen in the present case as well.

Urachal leiomyomas can mimic solid ovarian tumors or pedunculated uterine myomas.^[2,10] Benign urachal neoplasms can also mimic urachal malignancy. The radiological differential diagnosis also include adenocarcinoma of non-urachal origin, transitional cell carcinoma, infected urachal remnants and metastasis originating from primary lesions of colon, prostate or female genital tract.^[4,7] Because of lack of specificity of CT and ultrasonography in the differential diagnosis of solid urachal masses, a definitive pathological diagnosis is required to optimize the surgical approach and preclude unnecessary radical surgery.^[4]

Management of urachal lesions is surgical. The most accepted approach is resection of the urachus throughout its entire length. Minimally invasive surgical techniques are being currently advocated. Navarrete *et al.* have demonstrated the laparoscopic excision of urachal lesions via three 10-mm ports. The first port, through which pneumoperitoneum is performed is placed on the right hemi abdomen, at the umbilicus and the lateral border of the rectus muscle. The next ports are placed

on the epigastrium and the line joining the umbilical cicatrix with the right antero-superior iliacus spina. This minimally invasive surgical technique is safe and efficient, giving optimal post-operative results.^[11] For benign urachal lesions extending into the dome of the urinary bladder, excision of the urachus along with partial cystectomy is advocated. This can be done via open surgery, or by laparoscopic abdominal exploration.^[12] Primary treatment of potentially localized urachal carcinoma includes wide local excision of the urachus, umbilicus, and surrounding soft tissue combined with partial or radical cystectomy and bilateral pelvic lymphadenectomy. Although radical cystectomy has historically been advocated, several studies have demonstrated long-term survival with extended partial cystectomy including en bloc removal of the umbilicus, urachal tumor mass, the entire urachal ligament, and bladder dome.^[13,14] Chemotherapy and radiation for urachal adenocarcinoma have resulted in minimal responses with no definitive improvement in survival.^[13] Siefker-Radtke *et al.* reported the results of a retrospective review of 42 patients treated at the MD Anderson Cancer Center. Among the 26 patients who developed metastases, only 4 had significant responses to chemotherapy, and of the 9 patients who received chemotherapy with 5-FU or cisplatin containing regimens, 3 responded.^[13,15] Despite measurable responses to treatment, tumors often recur and the majority of patients die within 2 years of diagnosis.^[13]

To conclude, due to their rarity, these urachal remnant diseases are frequently misdiagnosed and confused with a wide spectrum of midline intra-abdominal or pelvic disorders.^[3] Understanding the embryological basis of these urachal disorders and their imaging features coupled with histopathological examination is crucial for the correct diagnosis and management.^[4]

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