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Case Report

Urachal mucinous adenocarcinoma in the pelvic wall mimicking endometriosis

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ABSTRACT

We report the case of a 30-year-old woman who complained of a painful palpable mass. Magnetic resonance imaging revealed an ill-defined mass approximately 8 cm in diameter with internal microcystic components. The mass diffusely involved the subcutaneous tissues, the muscles of the pelvic wall, and urinary bladder via a postoperative scar and resembled endometriosis. The histopathologic diagnosis was mucinous adenocarcinoma arisen from the urachal remnant. This is a very rare case of urachal adenocarcinoma arising mainly in the pelvic wall and mimicking endometriosis on MRI.

Introduction

Urachal neoplasms differ histologically from other bladder neoplasms on the basis that urachal remnants resemble the intestinal epithelium rather than the urothelium. Malignant urachal neoplasms account for less than 1% (0.35%-0.7%) of all bladder cancers [1,2]. The most common histologic subtype is adenocarcinoma [1,3]. Urachal adenocarcinoma is an uncommon malignancy accounting for 20%-39% of adenocarcinomas of the bladder [4,5]. Patients usually present at an advanced stage, and thus the prognosis is poor. Differentiating urachal adenocarcinoma from primary tumors of the surrounding structures and from metastasis is often

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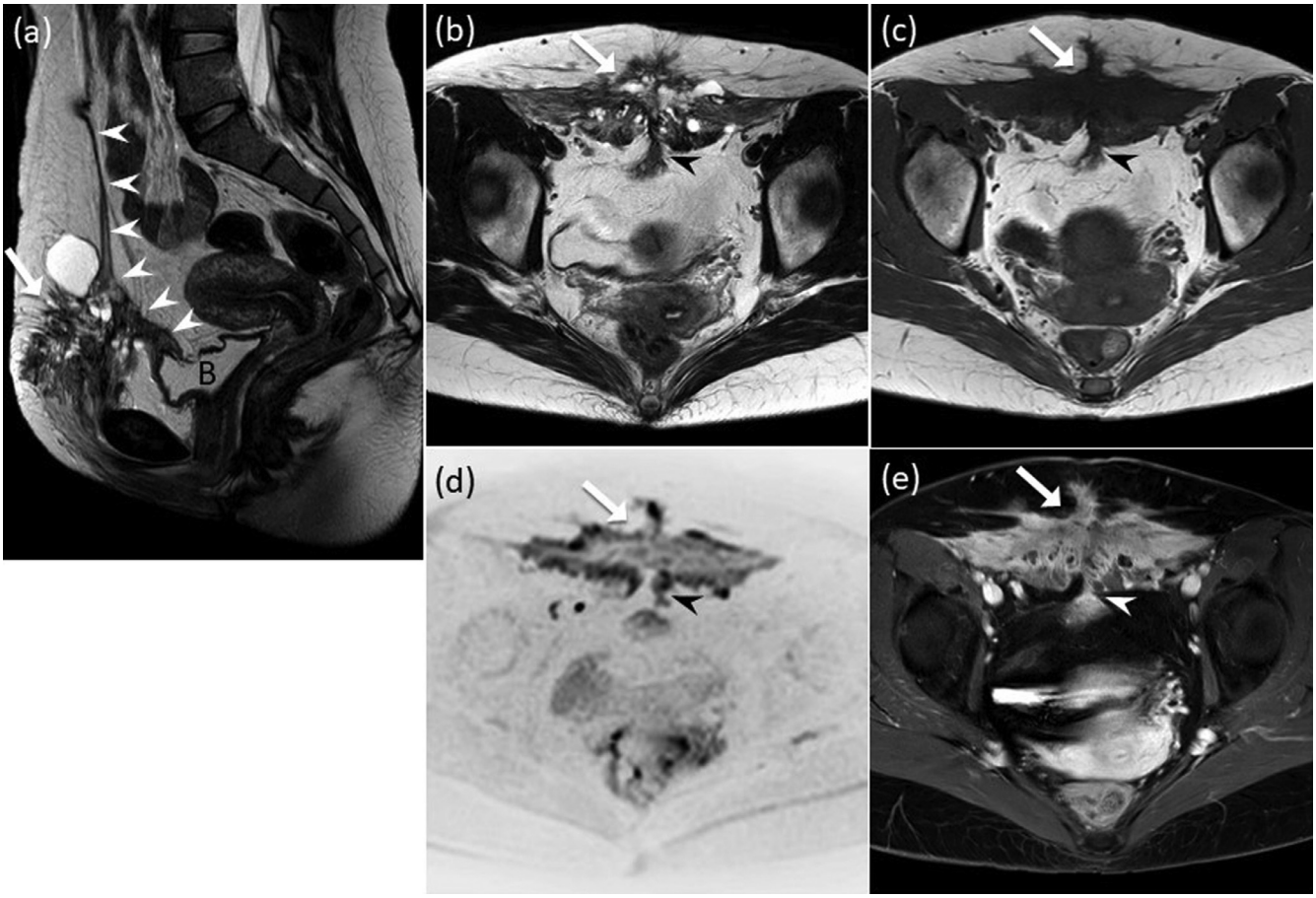

Fig. 1 – Magnetic resonance images of the mass in the pelvic wall. (a) Sagittal T2-weighted image shows an ill-defined mass in the pelvic wall (arrow). The mass shows low signal intensities with small high signal foci. Retrospectively, we considered that the linear structure (white arrow heads) continuous from the tumor to the bladder (B) and the umbilicus indicating the urachal remnant. (b) On axial T2-weighted image, the mass (arrow) diffusely involves the subcutaneous tissues and muscles of the pelvic wall. The mass communicates with the urinary bladder via the postoperative scar (black arrow head), which we retrospectively considered as the urachal remnant. (c) There was no internal high signal on T1-weighted images to indicate hemorrhage. (d) The mass (arrow) and urachal remnant (black arrow head) show mild restriction on diffusion weighted image with a b value of 1000s/mm^2. (e) The mass (arrow) and urachal remnant (white arrow head) are well enhanced on contrast-enhanced fat-saturated T1-weighted image.

difficult. We here report magnetic resonance (MR) imaging findings for a 30-year-old woman with urachal mucinous adenocarcinoma mimicking scar endometriosis.

Case report

A 30-year-old woman was referred to our hospital with a complaint of a painful palpable mass in the anterior pelvic wall. The mass had been identified a few years earlier. The relationship between pain and menstruation was unclear and she had no urinary symptoms. The hematologic and blood chemistry tests findings were normal and blood tumor markers were not elevated. She had a history of trans-abdominal enucleation of a serous cystadenoma of the right ovary by laparotomy, but no cesarean section.

MR imaging of the pelvis revealed an ill-defined diffusely infiltrating mass, approximately 8 cm in diameter, in the anterior pelvic wall. The mass showed low intensities with small high signal foci on T2-weighted images (Fig. 1a and b). T1-weighted image did not show high signal foci, which would indicate hemorrhage (Fig. 1c). Diffusion-weighted images showed mild restriction of the mass (Fig. 1d). The mass was well enhanced on the contrast-enhanced images (Fig. 1e). Small high signal foci on the T2-weighted images did not enhance and were considered cystic components. The mass diffusely involved the subcutaneous tissues and infiltrated into the bilateral rectus abdominis muscles and other pelvic wall muscles, communicating with the urinary bladder via the postoperative scar. Calcification was not detected on computed tomography (CT).

While there was no evidence of endometriosis in the pelvis, scar endometriosis of the pelvic wall was mostly suspected from the MR imaging findings and her past history (at the

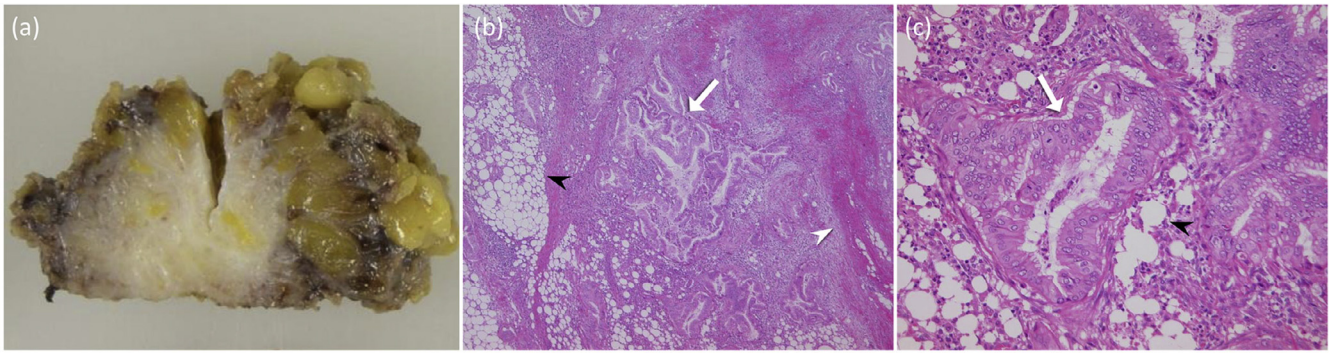

Fig. 2 – (a) Fixed sample of the partial excision shows a grayish white tumor and yellowish subcutaneous fat around it. (b) Microscopic examination (hematoxylin–eosin staining, original magnification, x 40) shows the tumor was composed of intestinal epithelium-like columnar epithelium with mucin production. Tumor cell glandules (arrow) were diffusely infiltrating subcutaneous fat (black arrow head) with fibrosis (white arrow head). (c) Microscopic examination (hematoxylin–eosin staining, original magnification, x 200) shows tumor cells with nuclear atypia and fission accompanied by clear cytoplasm (arrow).

time of imaging diagnosis, we did not know the reason for the unilateral ovarian enucleation). The differential diagnosis at the time included adenosarcoma and desmoid tumor. Biopsy was not easy because of the tumor's location in the deeper layers of the pelvic wall, and therefore, hormonal therapy (gonadotrophin-releasing hormone analogue) was performed. During the therapy, ultrasound was used for follow-up and reduction effect could not be confirmed. Eight months later, MR imaging revealed an increase in the size of the mass, and partial resection was performed.

Histologically, the mass was diffusely infiltrating adenocarcinoma with cystic components and fibrosis (Fig. 2a). Microscopically, the tumor was composed of intestinal epithelium-like columnar epithelium with mucin production (Fig. 2b and c). By immunohistochemistry, the tumor cells were positive for cytokeratin 7, cytokeratin 20, and CDX-28 (weak), and negative for estrogen receptor. No continuity with the intestine or ovaries was seen on imaging modality so the final diagnosis was mucinous adenocarcinoma originated from the urachal remnant.

FDG positron emission tomography-CT of the chest, abdomen, and pelvis did not reveal any distant metastasis. Complete resection seemed to be difficult because of the tumor's diffuse infiltration and she moved to another hospital for second opinion. Another 6 months later, she came back to our hospital. The mass developed more extensively and infiltrated to the right pubic bone (Fig. 3). An elevation of tumor marker CA19-9 (199 U/mL) also appeared. Chemotherapy started, but there was not much effect and the mass increased gradually.

Discussion

Although the normal urachus is most commonly lined with transitional epithelium, urachal carcinomas predominantly manifest as adenocarcinomas, which are lined with mucus-secreting epithelial cells, probably owing to metaplasia of the urachal mucosa into the columnar epithelium with subsequent malignant transformation [4,6].

A recent population-based study revealed that 82.4% of the malignant urachal tumors were adenocarcinomas and mucinous adenocarcinoma was the most common (42.9%). Malignant urachal tumors were predominantly seen in white (77.6%) and male patients (59%) aged older than 50 years (median age: 59, interquartile range: 46-71) and were usually low grade (37.9%) but diagnosed at advanced stages. The grade and disease stages were independently associated with cancer-specific mortality. The median overall survival time was 57 months, and the median cancer-specific survival time, 105 months. The 5-year overall survival and cancer-specific survival rates were 51% and 57%, respectively. The mortality rates did not differ between patients who underwent partial cystectomy and those who underwent radical cystectomy/exenteration, even after controlling for the tumor stage [1]. Although the most common symptoms of urachal cancers are hematuria, pain, and mass, urachal tumors are typically silent because of their extraperitoneal location. Consequently, late symptom presentation, early local invasion, and distal metastasis lead to their poor prognosis. In addition, their response to radiotherapy and chemotherapy is modest [3].

On imaging, urachal mucinous adenocarcinoma appears to be a mixed solid and cystic mass [7], although sometimes the tumor appears solid. The cystic components commonly seen in these tumors are mucin producing and are focally hyperintense on T2-weighted imaging and highly suggestive of adenocarcinoma. The solid components are isointense relative to soft tissue on T1-weighted images and are enhanced after contrast administration [8]. Peripheral calcification is also commonly seen on CT scans [8,9]. In most cases, the bulk of the tumor can be seen in the midline outside the lumen of the bladder with a predilection for the dome of the bladder. But, in some cases, tumor arises from the urachal remnant of the abdominal wall [10]. Diagnosis of urachal carcinoma is usually confirmed by cystoscopy and biopsy [8].

Scar endometriosis often infiltrates the deeper layers of the pelvic or abdominal wall, commonly the rectus abdominis

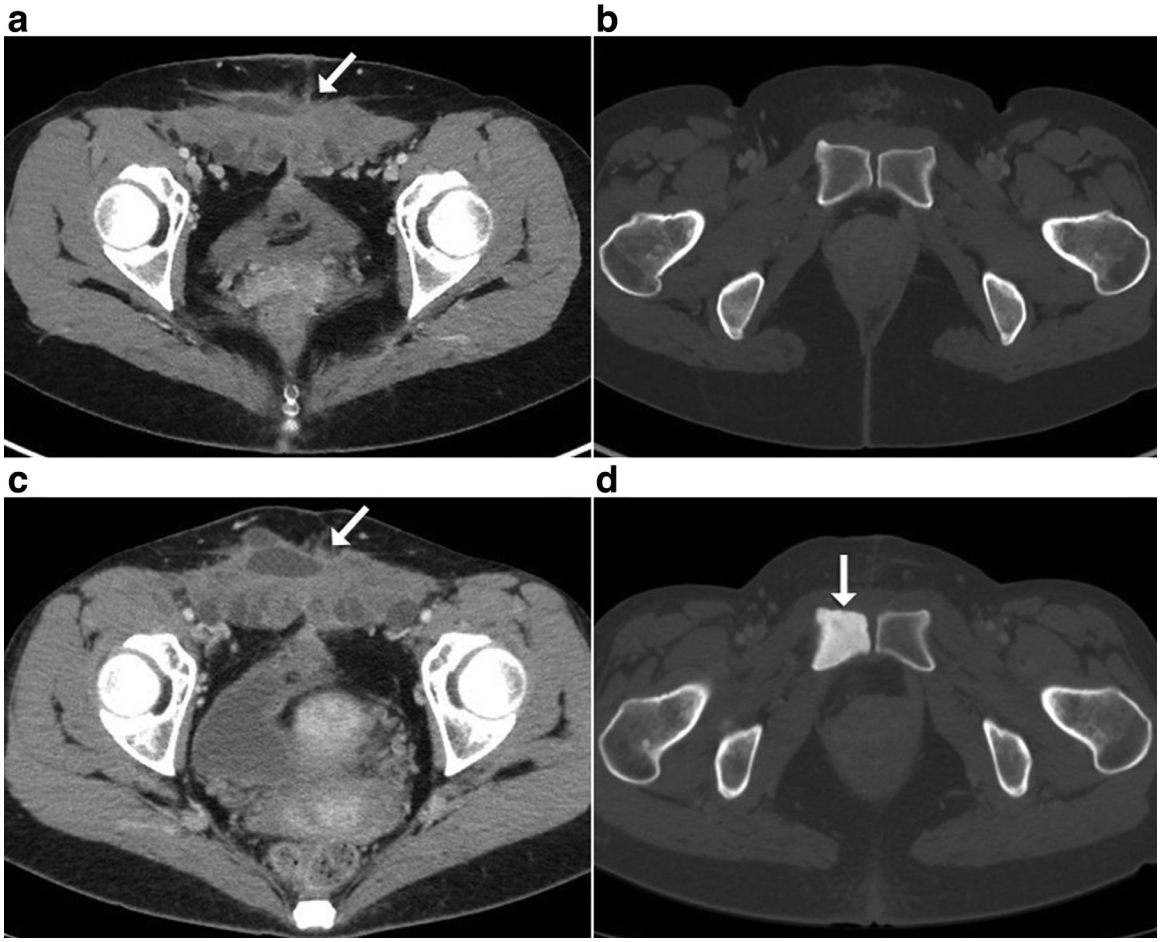

Fig. 3 – Computed tomography (CT) images of the mass in the pelvic wall. (a, b) At the time of a pathologic diagnosis confirmed. (c, d) Eight months later. (a, c) The mass (arrows) increases in size and develops more extensively with enlargement of the internal cystic components. (b, d) The mass has reached to the bilateral pubic bones and sclerosing change of the right pubic bone (arrow) indicating infiltration of the tumor has appeared.

muscle. Endometrial implants are categorized as cystic, solid, and mixed solid and cystic. The imaging findings of scar endometriosis vary greatly depending on the phase of the patient's menstrual cycle, the amount of fibrosis, glandular elements, bleeding, and inflammation. Sometimes, the signal intensity of scar endometriosis may be as low as that of muscle on T1- and T2-weighted images because of the fibrotic and hemosiderotic components. And usually, at least a portion of the lesion shows strong enhancement on contrast-enhanced images. Many patients with scar endometriosis do not have peritoneal endometriosis; thus transport of endometrial stem cells into pelvic wall incisions at the time of uterine surgery, followed by proliferation at the incision site, is the most plausible theory of scar endometriosis [11].

In our case, the mass showed very low signal intensities with internal microcystic components on T2-weighted images, which existed mainly in the postincision site of the anterior pelvic wall, and it affected this young patient with pain. Thus, we suspected scar endometriosis in the first. Actually, mucinous adenocarcinoma is infiltrated with fibrosis and cysts, and the MR imaging findings closely resembled endometriosis. The urachal remnant could not be identified as a separate structure even by pathologic investigation in our case; however, it was inferred by the imaging findings that a linear structure continuous from the tumor to the bladder (Fig. 1, arrow heads) was the urachal remnant. Probably, the urachal carcinoma easily infiltrated the subcutaneous tissues in the pelvic wall via the postoperative scar or arose from the urachal remnant that had adhered to the incision site.

To our knowledge, this is the first reported case of urachal adenocarcinoma mainly involved the subcutaneous tissues in the pelvic wall and mimicking endometriosis on MR imaging. Radiologists and gynecologists should keep this disease in mind as a possible pitfall in the differential diagnosis of ectopic endometriosis, and careful attention is required to the continuity between the bladder.

Disclosure

The authors have nothing to disclose.

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