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Intrapelvic sclerosing epithelioid fibrosarcoma: a case report

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Summary

Sclerosing epithelioid fibrosarcoma (SEF) is a rare deep-seated tumor, that typically develops in the lower extremity, limb girdle, and trunk. This tumor is clinically aggressive, with a poor prognosis. The authors present an extremely rare case of SEF developing in the pelvis. The patient was a 75-year-old woman presenting with a solid mass located adjacent to the uterus in the left adnexal region on vaginal ultrasonography. MRI showed a lobulated solid mass exhibiting hypointensity on T1- and T2-weighted imaging. The tumor also showed heterogeneous signal intensity on diffusion-weighted imaging, and heterogeneous enhancement on dynamic study, and both features were distinct in the tumor's peripheral zone. These MRI features were correlated with more hypercellularity in the peripheral zone on histology. To date, few reports have focused on the MRI features of SEF. Diffusion-weighted and contrast-enhanced MRI may provide more useful information for the diagnosis of SEF located in uncommon sites.

Key words: Fibrosarcoma; Sclerosing epithelioid fibrosarcoma; Pelvic mass; Magnetic resonance imaging; Immunohistochemistry.

Introduction

Sclerosing epithelioid fibrosarcoma (SEF) is a rare distinctive variant of fibrosarcoma, first described by Meis-Kindblom *et al.* in 1995 [1]. Its clinically aggressive behavior is reported with a recurrence rate of 40% to 50%, a metastasis rate of 40% to 80%, and a mortality rate of 31% to 57% from the disease [1-3]. SEF is a deep-seated tumor, most commonly occurring in the lower extremity, limb girdle, and trunk. There are few reports of the tumor arising in the retroperitoneum and abdominal and pelvic cavity [1, 4-6]. Because of its rarity, SEF may pose a diagnostic challenge, especially in cases of a tumor located in these uncommon sites. Herein, the authors report an extremely rare case of SEF occurring in the pelvis and focus on its histologic characteristics and MRI features. To date, few reports have focused on the MRI features of SEF [7].

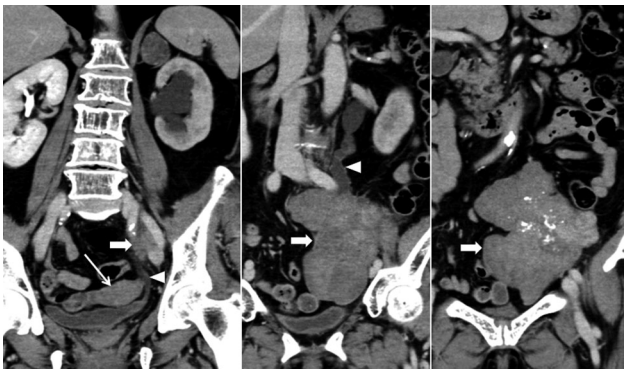

Figure 1. — Coronal images of contrast-enhanced CT demonstrate an irregular-shaped solid mass containing scattered granular calcification (short arrow) that involves the left ureter (arrowhead) and led to left hydronephrosis. The uterus (long arrow) is absent of tumor invasion.

Case Report

A 75-year-old Japanese woman, gravida 2, para 2, was diagnosed to have left hydronephrosis by screening abdominal ultrasonography. CT showed a solid mass in the left of the intrapelvic cavity. The patient had no reports of signs or symptoms of the conditions. She had a medical history of hypertension, impaired glucose tolerance, and a left adrenal adenoma that had been regularly followed up by an internist.

The woman was referred to the present hospital. Vaginal ultrasonography showed a solid mass located adjacent to the uterus in the left adnexal region. Contrast-enhanced CT showed an irregular-shaped solid mass (13×12×8.5 cm in size) containing scattered granular calcification that involved the left ureter and led to left hydronephrosis (Figure 1). Apparent lymph node swelling and metastatic disease were not observed.

MRI showed a lobulated solid mass exhibiting hypointensity on T1-weighted imaging (T1WI) (Figure 2A) and hypointensity containing granular or small irregular-shaped foci of intermediate signal on T2-weighted imaging (T2WI) (Figure 2B). The tumor also showed heterogeneous signal intensity on diffusion-weighted imaging (DWI), especially hyperintensity in the tumor's peripheral zone (Figure 2C). Dynamic study showed heteroge-

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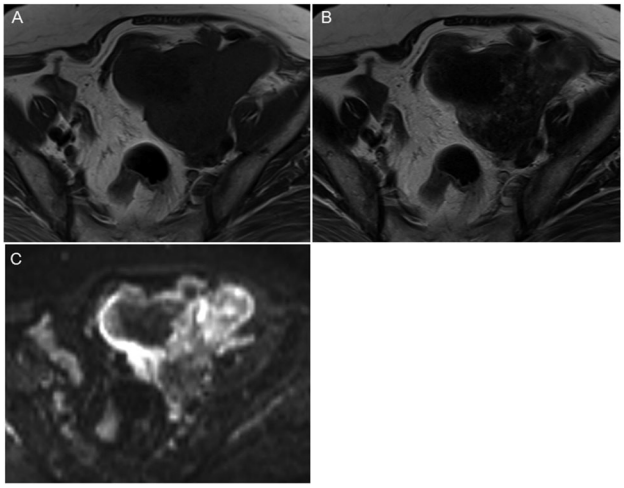

Figure 2. — Axial images of MRI of the pelvis demonstrate a lobulated solid mass showing hypointensity on T1-weighted imaging (A). Hypointensity containing granular or small irregular-shaped foci of intermediate signal on T2-weighted imaging (B). Heterogeneous signal intensity on diffusion-weighted imaging, especially hyperintensity in the tumor's peripheral zone (C).

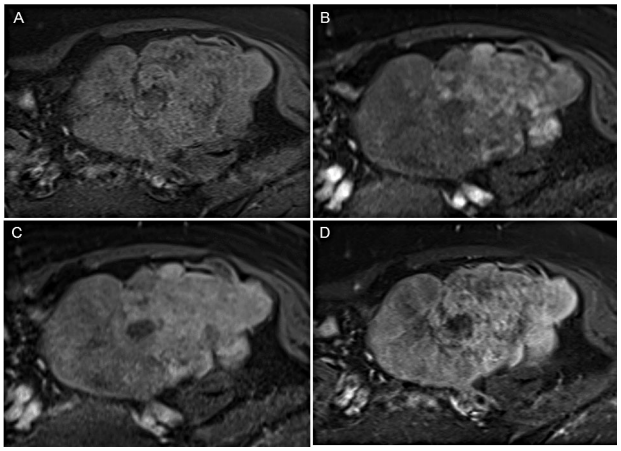

Figure 3. — Axial images of dynamic study demonstrate heterogeneous enhancement in the early phase (A [pre-contrast] to B), and gradually increased enhancement (C), especially marked enhancement in the peripheral zone (D).

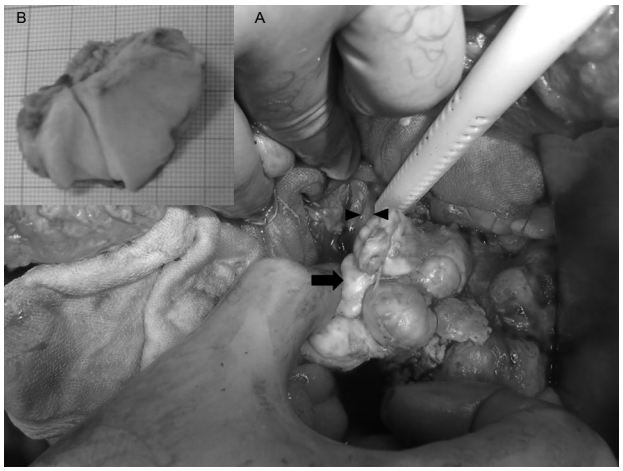

Figure 4. — (A) Intraoperative findings demonstate multinodular and firm tumor (arrow) located posteriorly to the left ovary and fallopian tube, involving the left ureter (arrowhead). (B) The cut surface of the tumor is grayish white.

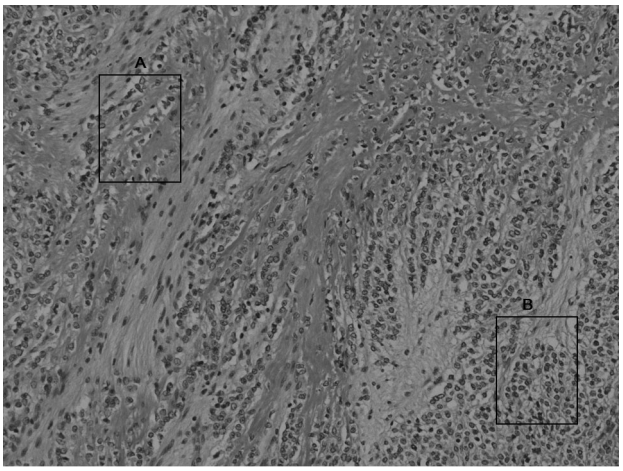

Figure 5. — Histologic findings demonstrate relatively uniform tumor cells arranged in cords and nests, surrounded by prominent fibrous tissues and densely hyalinized collagen in the central zone of the tumor (A). In the peripheral zone of the tumor, hypercellularity and scant fibrous tissues are observed (B) (H&E stain, ×20).

neous enhancement in the early phase (Figure 3B), and gradually increased enhancement (Figure 3C), especially marked enhancement in the peripheral zone (Figure 3D). MRI also showed an atrophic uterus without tumor invasion.

Gastroscopy and colonoscopy revealed no abnormal findings. Cervical cytologic smear yielded a negative result. The levels of serum tumor markers for carbohydrate antigen (CA) 125, CA19-9, carcinoembryonic antigen, α-fetoprotein, and lactate dehydrogenase were within the normal limits, but serum inflammatory response and slightly renal dysfunction were observed with the following findings: C-reactive protein, 7.09 mg/dl; blood urea nitrogen, 23.0 mg/dl; and creatinine, 0.83 mg/dl. The preoperative clinical diagnosis was an ovarian or fallopian tube carcinoma.

On laparotomy, the tumor was located posteriorly to the left ovary and fallopian tube, involved the left ureter, and extended to the left pelvic wall. The bilateral ovaries and fallopian tubes and uterus were not remarkable. This tumor was multinodular and firm, and it formed extensive adhesion to a sigmoid colon and the surrounding tissue with a tendency to bleed easily (Figure 4A). Therefore, the authors could not surgically resect this tumor, but they performed large biopsies and inserted a ureteral stent catheter into the left ureter.

The tumor's cut surface was gray-white (Figure 4B). Histologically, the tumor was composed of relatively uniform cells

arranged in cords and nests, surrounded by prominent fibrous tissues that varied from strands to insular shape. Individual tumor cells were demarcated from densely hyalinized collagen in the tumor's central zone (Figure 5A). In the tumor's peripheral zone, hypercellularity and scant fibrous tissues were observed (Figure 5B). The tumor cells had small oval nuclei and clear to pale eosinophilic cytoplasm. Mitotic figures were infrequent.

On immunohistochemistry, the tumor cells were positive for MUC4, desmin, and vimentin, but negative for broad-spectrum keratins, S100, neurofilament, HHF35, and MIC2. MIB-1 labeling index was 5%. The final pathologic diagnosis was SEF.

The patient and her family did not desire any additional therapy. Although the tumor increased to 14×20×14 cm from 13×12×8.5 cm on CT imaging, no evidence of metastasis was detected within a 24-month postoperative period.

Discussion

SEF is a rare clinicopathologically distinct variant of fibrosarcoma that typically involves deep soft tissues of the lower extremities, limb girdle, trunk, the upper extremities, and the head and neck. Uncommon sites of its origin were reported as the retroperitoneum, kidney, pancreas, cecum, omentum, mesentery, pelvic cavity, and ovary [1, 4-6].

Histologically, SEF is characterized by low-grade tumor features: a proliferation of small to medium-sized epithelioid cells arranged in cords and nests in a densely sclerotic hyalinized stroma. Tumor cells have round to oval bland nuclei and variable amounts of clear or pale eosinophilic cytoplasm. Mitotic activities are rare. Immunohistochemically, MUC4 is a sensitive and relatively specific marker for SEF [8]. The tumor cells are typically negative for keratins, CD34, smooth muscle actin, and desmin [5, 8]. A subset of SEF is related to low-grade fibromyxoid sarcoma [2,8]. Recently, fluorescence *in situ* hybridization studies showed the characteristic *EWSR1-CREB3L1* gene fusion in SEF [6].

SEF's preoperative estimation is challenging, especially in those affecting uncommon sites. MRI is considered as a very useful tool for detecting and differentiating intrapelvic lesions. Christensen *et al.* demonstrated that SEF showed a zonal architecture on MRI [9]. The tumor was composed of a large central stellate core showing very low signal intensity on both T1WI and T2WI and a peripheral zone showing intermediate signal intensity on T1WI and intermediate to high signal intensity on T2WI. The former was correlated with decreased cellularity and dense hyalinized collagen background matrix, and the latter was correlated with more hypercellularity and more nuclear pleomorphism on histology.

The present case showed features similar to those of the reported case, but more heterogeneous features on T2WI. Furthermore, this case showed characteristic features of a heterogeneous signal intensity on DWI and heterogeneous enhancement on dynamic study, and both features were also distinct in the tumor's peripheral zone. To the best of the present authors' knowledge, there are few studies on contrast enhancement and diffusion-weighted MRI for SEF.

The differential diagnoses of the intrapelvic SEF on MRI included uterine subserosal leiomyoma, uterine leiomyosarcoma, ovarian fibroma/thecoma, metastatic ovarian tumor, neurofibroma, desmoid-type fibromatosis, and soft-tissue sarcoma containing abundant collagenous fibers. Uterine leiomyoma typically shows low to intermediate signal intensity compared to normal myometrium on T1WI and low signal intensity on T2WI. However, degenerated leiomyomas include variable signal intensity on T2WI and variable enhancement with contrast administration. Tanaka *et al.* demonstrated that leiomyosarcomas are highly suspected, when more than 50% of the tumor shows high signal intensity on T2WI, any small high signal areas are seen within the tumor on T1WI, and some unenhanced pocket-like areas are seen after administration of contrast materials [9]. Detection of a pedicle and flow voids of the tumor toward the uterus is helpful for the diagnosis of uterine tumors.

Chung *et al.* demonstrated that ovarian fibrothecomas typically showed homogenously low to intermediate signal intensity on T1WI and low signal intensity on T2WI. However, they often showed atypical imaging features that mimic malignancy. They noted that low signal intensity on DWI and mild enhancement with contrast enhancement may help us consider the fibrothecoma as a benign tumor [10]. Detection of normal ovaries on T2WI is also helpful to exclude ovarian tumors.

Kransdorf *et al.* reviewed the MRI images of 112 soft-tissue masses of various causes and then concluded that conventional MRI could not reliably distinguish between benign and malignant soft-tissue tumors [11].

SEF is a clinically aggressive tumor with a high recurrence rate and poor prognosis, although it is a histologically low-grade sarcoma. Optimal treatment regimens have not yet been determined because of its rarity. Surgical resection is currently the only appropriate treatment [3, 5]. Therefore, early and accurate diagnosis is necessary.

Conclusion

MRI is a very useful diagnostic tool. Clinicians and radiologists should consider the possibility of SEF in a case of solid mass showing a characteristic zonal architecture on MRI. Contrast-enhanced and diffusion-weighted MRI may provide more useful information for the diagnosis of SEF, especially located in unexpected sites. Because of limited available data, additional clinical data are necessary for an accurate diagnosis of SEF originating from uncommon sites.

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