Variable Distribution of Pseudolobules in Ovarian Sclerosing Stromal Tumors: Utility of Diffusion-weighted Imaging for Differential Diagnosis

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Clinical Image

Sclerosing stromal tumors (SSTs) are rare benign sex cord–stromal tumors of the ovary. Distinguishing SST from other malignant ovarian tumors is crucial because SSTs affect adolescents and young adults (AYA) who require fertility preservation. Here, we present a case of SST with characteristic imaging findings and its correlating pathological findings.

A 17-year-old girl with irregular menstruation visited our institution. No abnormalities were identified by physical and laboratory evaluations. T2-weighted magnetic resonance (MR) images (T2WI; Fig. 1a) revealed a relatively hypointense solid mass affecting the left adnexa. This mass included multiple island-shaped areas surrounded by hypointense rims. These areas were indistinct on T1-weighted images (Fig. 1b). No signal voids were observed within or around the tumor. Compared to endometrium, islands-shaped areas appeared significantly hyperintense on diffusion-weighted imaging (DWI) (Fig. 1c), and restricted diffusion on apparent diffusion coefficient (ADC) map (Fig. 1d). These areas were as strongly enhanced as myometrium from the early phase to the equivalent phase of dynamic contrast enhancement (DCE) (Fig. 1e). Therefore, we speculated these island-shaped area might correspond to pseudolobules, a pathological hallmark of the SST. The preoperative imaging diagnosis was SST.

Macroscopically, the resected left ovarian tumor showed a multilobulated yellow cut surface. Histologically, cellular pseudolobules were separated by dense fibrous stroma (Fig. 2a). Dilated vessels were frequently observed in cellular areas (Fig. 2b). Immunohistochemically, tumor cells were α-inhibin-positive (Fig. 2c). The pathological diagnosis was SST.

SST predominantly affects AYA in whom the most common symptom is irregular menstruation. Pathologically, SST is characterized by a pseudolobular structure comprising hypervascular cellular areas separated by edematous or fibrous hypocellular areas. Hypointense nodules against hyperintense stroma on T2WI and striking enhancement have been reported as characteristic MR findings of SST.1 Although the stroma was expected to exhibit marked hyperintensity on T2WI, it was nearly isointense to the pseudolobules in our case. Rich collagen fibers within the background stroma might decrease the background signal and reduce the distinct contour of the pseudolobules. Pseudolobules comprised abundant tumor cells with hypervascularity, whereas the stroma comprised fibrous areas with low cellularity. We could thus recognize pseudolobules from the striking enhancement and restricted diffusion areas on MR. Matsubayashi et al. reported early peripheral tumor enhancement with centripetal progression on DCE2 as the pseudolobules were typically located at the tumor margin. However, in our case, the pseudolobules were scattered throughout the tumor and appeared as island-shaped areas with marked enhancement. These findings suggest wide variation in the distribution of pseudolobules and extent of stromal edema and fibrosis. DWI may be a promising technique for distinguishing pseudolobules with abundant tumor cells from edematous or collagenous background. Therefore, we speculate there are variety of distribution of pseudolobules in ovarian SSTs. So we carefully differentiate metastatic ovarian tumors from SSTs, which may also contain scattered tumor nest with distinct margins.2,3

In conclusion, regardless of the distribution of pseudolobulation and extent of edematous or fibrous stroma, DWI and DCE are useful for visualizing pseudolobules with abundant tumor cells and can facilitate the diagnosis of SST.

Conflicts of Interest

The authors declare that they have no conflicts of interest.
Fig 1. Magnetic resonance (MR) images of a 17-year-old girl with a sclerosing stromal tumor. Axial T₂-weighted imaging (repetition time [TR]/echo time [TE]: 3952/83.34 ms) reveals a solid non-invasive mass (arrows) with island-shaped areas of intermediate signal intensity surrounded by hypointense rims (arrowheads) (a). The mass shows low signal intensity on an axial T₁-weighted image (TR/TE: 220/4.78 ms) (b). Axial diffusion-weighted imaging (TR/TE: 5400/87 ms, b = 1000 s/mm²) shows island-shaped areas with significant hyperintensity (c). Diffusion coefficient (ADC) mapping demonstrates islands of hypointensity relative to the background stroma. ADC values for the hypointense islands and background stroma are 1.06 × 10⁻³ mm²/s and 1.69 × 10⁻³ mm²/s, respectively (d). During the early (50-s) phase of dynamic contrast enhancement, island-shaped areas are strikingly enhanced, whereas the background stroma is weakly enhanced (e). U, uterus.

Fig 2. Loupe image of the tumor shows pseudolobulation characterized by cellular areas separated by dense fibrous stroma with low cellularity. Few edematous changes are evident in the stroma (a: hematoxylin–eosin stain, loupe image). Cellular areas are composed of spindle cells and round cells resembling luteinized stromal cells. They have marked vascularity and prominent sclerosis around clusters of individual cells (b: hematoxylin–eosin stain, medium-power field). Tumor cells are positive for α-inhibin (c: high-power field).

References