Case Report

Ependymoma of the broad ligament mimicking an ovarian surface epithelial tumor

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\section*{Abstract}

We report a case of ependymoma of the broad ligament occurring in a 21-year-old woman. CT and MRI findings showed a 40-mm-diameter, well-demarcated cystic mass with a lobulated solid component in the right pelvis. The solid component showed heterogeneous intermediate signal intensity on T2-weighted image and prolonged mild contrast enhancement. The tumor was resected and confirmed as ependymoma based on the histologic findings along with its immunohistochemical profile. To our knowledge, this is the first report of an adnexal ependymoma describing the precise radiological characteristics that resembled those of borderline or malignant epithelial ovarian tumors.

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\section*{Introduction}

Primary extraneural ependymomas are rare tumors that arise in ectopic sites, including the posterior mediastinum, lung, ovary, paraovarian tissues and omentum [1–4]. Ependymoma of the broad ligament is extremely rare; only 5 cases have previously been reported, while there are 15 cases of ovarian ependymoma in the literature [3,5–8]. The detailed radiological features of these tumors, however, have not been reported. The present report describes a case of an ependymoma of the broad ligament, for which we could thoroughly review the imaging findings.

\section*{Case report}

A 21-year-old woman presented with a right pelvic mass. She had undergone a periodic medical examination and had been found to have a pelvic mass 2 years earlier. She had neither symptoms nor abnormal blood test including tumor makers of

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carcinoembryonic antigen (CEA), carbohydrate antigens CA19-9, CA125, and alfa-fetoprotein (AFP).

Radiological findings

Non-contrast and contrast-enhanced CT showed a 40-mm-diameter, well-demarcated low-attenuating mass with solid and cystic components in the right pelvis. The CT numbers of the solid component were 47 HU and 77 HU on noncontrast and contrast-enhanced CT, respectively (Fig. 1). There were neither calcifications nor gross fat attenuation. On MRI, the lesion was located adjacent to the right ovary without apparent “beak sign.” The cystic component showed iso-intensity on T1WI and prominent hyperintensity on T2WI (Fig. 2A and B). The solid component projecting into the cyst demonstrated a papillary structure with intermediate intensity and punctate foci of high intensity on T2WI (Fig. 2B). The solid component showed higher intensity than the ovarian stroma on diffusion-weighted image. The apparent diffusion coefficient (ADC) value ranged from 1.0 to 1.2 × 10⁻³ mm²/s (Fig. 2C and D). On dynamic contrast-enhanced T1WI, the solid component showed heterogeneous and prolonged enhancement (Fig. 2E). The differential diagnoses were made as ovarian serous borderline tumor, serous adenocarcinoma, and endometrial borderline tumor based upon these radiological findings.

Operative findings

A well-mobile 40-mm-sized mass was found beside the fimbria abutting the fallopian tube and right ovary (Fig. 3). The right ovary was intact. The tumor, fallepiant tube, and a part of the right ovary were resected. The content of the cystic component was serous.

Pathologic findings

The mass was a cystic tumor with a smooth internal surface including a yellowish polyp-like component (Fig. 4A). Microscopically, the solid component showed papillary structure with a lobulated surface on H&E stain (Fig. 4B). True rosettes composed of tumor cells surrounding a lumen as well as perivascular pseudorosettes formed by tumor cells radially surrounding blood vessels, which featured a non-nucleated zone around the vessels, were observed (Fig. 4C and D). No nuclear atypia was found. Immunohistochemistry showed strong positivity for glial fibrillary acidic protein (GFAP) in the cytoplasm. The neoplastic cells were also positive for vimentin, chromogranin, S-100, and Wilms’ tumor 1 and negative for cytokeratin AE1/AE3, neurofilament, and inhibin (Fig. 4E-G). MIB-1 was positive in less than 5% of the tumor cells. Based on these immunohistologic findings, the tumor was comprehensively diagnosed as an ependymoma.

Discussion

Ependymomas account for up to 5% of neuroepithelial neoplasms that arise from the lining of the ventricles of the brain or the central canal of the spinal cord. These tumors can occur in any age group but are more common in younger patients. Those that occur in the posterior fossa are more common in children, whereas the mean age for supratentorial lesions is 18-24 years [9]. They rarely metastasize or arise ectopically [10-12]. Histopathologic features include true rosette, perivascular pseudorosette, and GFAP-posivity on histochemical staining [9]. Microscopically, they tend to be well-circumscribed tumors with heterogeneous solid and cystic components [13]. While the recognizable 5 cases of ependymoma of the broad ligament reportedly presented as heterogeneous solid tumor with or without macroscopic cys-

Fig. 1 – Noncontrast CT (A) and contrast-enhanced CT (B) shows a 40-mm-diameter, well-demarcated low attenuating mass with a solid and a cystic component in the right pelvis. The CT numbers of the solid component are 47 HU and 77 HU on noncontrast and contrast-enhanced CT, respectively. There are neither calcifications nor gross fat-attenuation.
Fig. 2 – The cystic component is iso-intense on T1WI (A) and prominently hyper-intense on T2WI (B). The solid component that projected into the cyst demonstrates a papillary structure with intermediate intensity and punctate foci of high intensity on the T2WI (arrowhead). The lesion is located adjacent to the right ovary (arrow) without apparent “beak sign” between them. The solid component (arrowhead) shows higher intensity than the ovarian stroma on diffusion-weighted image (C). The ADC value ranges from 1.0 to 1.2 × 10⁻³ mm²/s (D). Dynamic contrast-enhanced T1WI shows heterogeneous and prolonged enhancement in the solid component (E, pre-contrast; F, arterial phase; G, equilibrium phase).
The tumor was found adjacent to the fimbria of the fallopian tube (A). The opened-up specimen shows a smooth internal surface of the cystic component and a yellowish polypoid solid component (B).

H&E stain shows papillary structures with a lobulated surface in the solid component (A, low power), true rosettes (B, high power), and perivascular pseudorosettes (C, high power). Immunohistochemical staining shows positive staining for GFAP (D) and vimentin (E), whereas negative for AE1/AE3 (F).

tic component like intracranial ependymomas [3,5–8], primary extraneural ependymomas have been reported to have a wider range of microscopic architecture distinct from intracranial ependymomas [12]. Although its histogenesis still remains unclear, one suggests that ependymomas of the broad ligament arise from totipotent germ cells in the teratoma similar to those in ovarian ependymomas which are classified as a neuroectodermal tumor in the monodermal teratoma [7].

Radiologically, intracranial ependymomas usually show heterogeneous signal intensity on MRI due to their various proportions of cystic and solid components with intracellular myxoid accumulation [14]. They demonstrate heterogeneous, weak to moderate enhancement [14]. Findings on diffusion-weighted images are variable. One study showed the mean minimum ADC value of ependymomas in the fourth ventricles was 1.11 ± 0.13 (SD) mm²/s [15]. The cystic component tends to be similar to the cerebrospinal fluid in signal intensity both on T1WI and T2WI, but it may not be completely suppressed on FLAIR images due to proteinaceous content [14]. On the other hand, radiological features of extraneural ependymomas have not been well-described to date due in part to their rarity. Kumari et al. [8] reported a case of ependymoma of the broad ligament as a heterogeneous solid tumor without cystic components on a contrast-enhanced CT study. In our case, the tumor was a well-circumscribed cystic lesion with a heterogeneous mural nodule with diffusion re-
striction that were also consistent with findings in borderline or malignant surface-epithelial tumors. One possible distinct radiological feature that might be able to discriminate this case from those epithelial tumors was the solid component that showed intermediate intensity with punctate foci of hyperintensity on T2WI, which might be due to its high-density lobulated papillary structure. In the context of these subtle imaging characteristics, teratomas with minimal fatty tissue component, in which adnexal ependymoma may be included, could be raised as another differential diagnosis. Nevertheless, the findings such as moderate and prolonged enhancement on dynamic contrast-enhanced T1WI and moderate diffusion restriction observed in this case were nonspecific and shared by many types of tumors including surface-epithelial ovarian tumors, making it difficult to narrow down differential diagnosis [16,17]. As with most cases of intracranial ependymomas, the tumor in our case did not show any evidence of necrosis nor hemorrhage on both CT and MRI [18]. The reported cases of ependymoma of the broad ligament were also proven to be free from necrosis or hemorrhage with an only exception of a case of 14-cm solid mass which demonstrated hemorrhaging and necrosis along with myxoid foci [7]. There were no calcification foci in our case as well as in all the reported cases of ependymoma of the broad ligament, while approximately 50% cases of intracranial ependymomas reportedly demonstrated coarse calcifications on CT [18].

Conclusion

We reported a case of ependymoma of the broad ligament that presented with diagnostic difficulty due to close resemblance to cystic epithelial ovarian tumors in radiological features. Nevertheless, MRI might provide signature findings that could be beneficial to differentiate these rare tumors from other relatively common adnexal tumors.

Patient consent statement

Written informed consent was obtained from the patient for publication of this case and any accompanying images.

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