CASE REPORT

A case of ependymoma arising from the peritoneum

Yasuko Yamamoto^{1,2} · Kazuhiro Takehara¹ · Shinichi Okame¹ · Masaaki Komatsu¹ · Yuko Shiroyama¹ · Takashi Yokoyama¹ · Shinichi Tanaka³ · Masakazu Sato³ · Rieko Nishimura³ · Norihiro Teramoto⁴

Received: 7 October 2015 / Accepted: 7 December 2015 / Published online: 15 December 2015
© The Japan Society of Clinical Oncology 2015

Abstract Ependymoma arising from the peritoneum is extremely rare. We present the case of a 23-year-old woman who underwent urgent laparoscopic surgery because of a pelvic mass and intraperitoneal bleeding. Although peritoneal carcinoma was suspected, pathological re-examination revealed ependymoma with a perivascular pseudorosette and positive for glial fibrillary acidic protein. Residual tumor extraction indicated that the ependymoma had developed from the peritoneum. This case highlights the need to consider ependymoma as a potential diagnosis in young women with suspected ovarian or peritoneal cancer.

Keywords Peritoneal tumor · Ependymoma · Female

Introduction

Ependymoma is defined as a tumor comprising neoplastic ependymal cells. It is a type of glioma that develops from central nervous tissue, such as the wall of the cerebral ventricle or central canal of the spinal cord. Ependymomas accounts for 3–5 % of all gliomas. Although the ovary and peritoneum have been reported as sites of origin for ependymomas, such cases are extremely rare and the clinical characteristics are unclear. We report an unusual case of ependymoma arising from the peritoneum.

Case report

A 23-year-old Japanese woman with no significant past medical or familial history presented to the emergency department of her previous hospital with complaints of abdominal pain, fever elevation, and vomiting. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed a pelvic mass and intraperitoneal bleeding (Fig. 1). The patient underwent urgent laparoscopic surgery. A clot and ruptured soft tumor mass were observed in the anterior wall of the uterus, pouch of Douglas, and great omentum. No macroscopically abnormal findings were observed in the bilateral adnexa. Pathological examination suggested adenocarcinoma, and she was therefore referred to our cancer center with suspicion of peritoneal cancer.

We detected masses around the right adnexa and pouch of Douglas based on the original MRI results, and abnormal shadows close to the liver margins were revealed by CT and fluorodeoxyglucose positron emission tomography–CT (Fig. 2a, b). Blood biochemistry results showed slightly elevated CA125 levels (53.2 U/ml; normal, 0–35 U/ml). Serum levels of CA19-9, carcinoembryonic

✉ Kazuhiro Takehara
katakehara@shikoku-cc.go.jp

¹ Department of Gynecologic Oncology, National Hospital Organization, Shikoku Cancer Center, Ko-160 Minami Umemoto, Matsuyama 791-0280, Japan

² Department of Obstetrics and Gynecology, Graduate School of Biomedical Science, Hiroshima University, Kasumi 1-2-3 Minami-ku, Hiroshima 734-8551, Japan

³ Department of Clinical Laboratory, National Hospital Organization, Shikoku Cancer Center, Ko-160 Minami Umemoto, Matsuyama 791-0280, Japan

⁴ Department of Pathology, National Hospital Organization, Shikoku Cancer Center, Ko-160 Minami Umemoto, Matsuyama 791-0280, Japan

Fig. 1 MRI at patient's previous first hospital visit. A solid lesion wrapped in a capsule-like structure was detected in the uterus ventral and dorsal aspects

Fig. 2 **a** CT showed a soft, abnormal shadow close against the liver S6 margins. **b** Postoperative PET-CT showed light fluorodeoxyglucose accumulation in these sites (SUVmax = 2.6)

antigen, and α -fetoprotein were within normal ranges. The previous diagnosis was suspicious of serous carcinoma arising from the peritoneum (Fig. 3). However, we suspected ependymoma based on the detection of a perivascular pseudorosette structure (Fig. 4) comprising cytoplasmic processes surrounding a small vessel by hematoxylin and eosin staining of tissue sections. Immunohistochemical staining was positive for glial fibrillary acidic protein (GFAP) in the cytoplasm of the tumor cells (Fig. 5), confirming the diagnosis of ectopic ependymoma. Immunohistochemistry also revealed

positivity for vimentin, CD56, S-100, WT-1, estrogen receptor (ER), and progesterone receptor (PgR) (Table 1). No central nervous system abnormality was detected by cephalic imaging study. We made a final diagnosis of ependymoma arising from the peritoneum. Debulking surgery was planned involving residual tumor enucleation, because the patient wanted to preserve her fertility potential. Dissemination throughout the pelvic floor and a grayish-white papillary mass stemming from the retroperitoneum on the underside of liver were noted during laparotomy (Fig. 6). Her bilateral ovaries were

Fig. 3 Histologic findings demonstrated similar morphology to serous carcinoma at the initial laparoscopic surgery (papillary growth and psammoma body). **a** Hematoxylin and eosin (HE) $\times 200$, and **b** HE $\times 40$

Fig. 4 Perivascular pseudorosette. **a** HE $\times 200$, and **b** HE $\times 400$

Fig. 5 Immunohistochemical findings indicating GFAP positivity ($\times 200$)

Table 1 Immunohistochemical staining

Antibody	
GFAP ^a	+
Vimentin	+
CD56	+
S-100	+
p53	–
WT-1	+
Keratin7	(–)/p(+)
Calretinin	–
ER	+
PR	+
MIB-1 index (%)	5–20 %

^a GFAP Glial fibrillary acidic protein

Fig. 6 **a** Laparotomy findings. The mass developed from the retroperitoneum on the underside of the liver. **b** Macroscopic appearance. The tumor was a grayish-white papillary mass

macroscopically normal, but some miliary peritoneal lesions remained in the peritoneal pouch of Douglas. We suggested adjuvant chemotherapy, but the patient refused. No tumor progression was detected at 10 months after surgery.

Discussion

Ependymoma arising from the peritoneum is extremely rare, with only 17 case reported in Japan since its first record by Kleinman et al. in 1984 [1]. Although ependymomas occurring outside the central nervous system were classified as ‘monodermal and highly specialized teratomas’ according to the previous WHO classification [2], they are currently classified under miscellaneous tumors and ‘tumors of the broad ligament and other uterine ligaments’ in the 4th edition [3]. No macroscopic neoplastic lesions were found in either ovary in the current case, and the tumor was determined to have arisen from the peritoneum.

Ependymomas developing from the abdominal cavity are most common in women of reproductive age (mean age 35 years), and there have been no reported occurrences in men [4]. Ependymomas comprise a nodular mass with a mix of solid and cystic parts, sometimes with bleeding and necrosis, based on imaging examinations. Specific imaging findings of ependymoma in abdominal cavity have not been reported. Although positron emission tomography/computed tomography (PET/CT) findings have not been reported, our case indicated low fluorodeoxyglucose (FDG) uptake at tumor. Ependymoma arising from the peritoneum may have a low maximum standardized uptake

values (SUVmax) level of PET/CT compared to common peritoneal cancer.

Tumor growth is relatively slow. Duggan et al. reported no correlation between mitotic rate and prognosis, and suggested that tumor diameter was associated with risk of metastasis and recurrence [4]. There are a few case reports of recurrence, however, in most of those cases, the time to recurrence is several years (0.5–30 years, median 4.75 years) [5]. Since late recurrence is also reported, careful long-term surveillance of the patients is required [3]. In terms of treatment, surgical resection is considered to be important, but the value of postoperative chemotherapy has not been established. Although there have been several reported cases of the use of chemotherapy with cisplatin, and combination chemotherapy with bleomycin, etoposide, and cisplatin (BEP). However, Takano et al. reported that these treatment preferences were based on germ cell tumors [5].

Some disseminated lesion remained after debulking surgery in the current case because of the patient’s wish to preserve her fecundity. However, although we performed conservative surgery, there is currently no evidence of her prospects for pregnancy, and if the residual tumor subsequently grows or metastasizes, more radical therapies may be required, despite the patient’s preferences.

The pathological findings of ectopic ependymoma are similar to those of ependymoma of the central nervous system. Jacket rosette, perivascular pseudorosette, and an ependymal canal are characteristic, together with GFAP positivity on immunohistochemistry. If the site of origin is in the central nervous tissue, the diagnosis of ependymoma is relatively straightforward; however, in the case of an intraperitoneal origin, the papillary structure may lead to its

misdiagnosis as a serous carcinoma or endometrioid carcinoma, and a clinical diagnosis of ovarian cancer or peritoneal carcinoma. Patient age, tumor-development pattern, and the clinical picture should be considered in differentiating between ependymoma and common ovarian or peritoneal cancer. The fact that ependymomas may originate from the ovary or peritoneum suggest that they may develop by the following means: (1) as an ectodermal ingredient of teratoma; (2) by neometaplasia of a tissue derived from the peritoneum and Mueller duct; and (3) from ectopic nervous system and fetal components (derived from primordial germ cells) remaining in the pelvis. Ependymoma of the central nervous system show infrequent ER and PgR expression, whereas ependymomas that develop from the ovary or peritoneum are highly positive for ER and/or PgR [6]. ER and PgR expression may thus differ according to the origin of the tumor cells from central nervous tissue or not [7]. A previous case report described a patient with an ER- and PgR-positive ovarian ependymoma who achieved stable disease after postoperative treatment with an aromatase inhibitor [8]. However, it is currently uncertain if hormone sensitivity is associated with a specific regimen, and if adnexectomy leads to improved survival.

Because ectopic ependymoma is very rare, it is difficult to establish this diagnosis before surgery. In the 17 previous cases reported in Japan, we were able to access the data for 11 cases, among which malignancy was confirmed preoperatively in nine, and ovarian cancer was suspected in eight cases [9–11]. Seven of the 11 patients were in their 20 s. A previous report suggested that strong chemotherapy may be administered unnecessarily when serous or endometrioid carcinoma is diagnosed pathologically [12].

In conclusion, although ectopic ependymoma is rare, it is important to consider it as a possible diagnosis in young women with suspected ovarian or peritoneal cancer. Further cases are needed to establish the clinical characteristics and optimal treatment strategies for this condition.

Acknowledgments This research was partly supported by the Practical Research for Innovative Cancer Control from Japan Agency for Medical Research and development, AMED.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Research involving human participants and/or animals This article does not contain any studies with human participants or animals performed by any of the authors.

Informed consent For this type of manuscript, formal consent is not required.

References

1. Kleinman GM, Young RH, Scully RE (1984) Ependymoma of the ovary: report of three cases. *Hum Pathol* 7:632–638
2. Tavassoli FA, Devilee P (2003) Pathology and genetics of tumors of the breast and female genital organs. Word Health Organization Classification of tumors, 3rd edn. IARC Press, Lyon
3. Kurman RJ, Carcangiu ML, Herrington CS et al (2014) WHO Classification of tumors of female reproductive organs. Word Health Organization Classification of tumors, 4th edn. IARC Press, Lyon
4. Duggan MA, Hugh J, Nation JG, Robertson DI et al (1989) Ependymoma of the uterosacral ligament. *Cancer* 64:2565–2571
5. Takano T, Akahira J, Moriya T et al (2005) Primary ependymoma of the ovary: a case report and literature review. *Int J Gynecol Cancer* 15:1138–1141
6. Idowu MO, Rosenblum MK, Wei XJ et al (2008) Ependymomas of the central nervous system and adult extra-axial ependymomas are morphologically and immunohistochemically distinct—a comparative study with assessment of ovarian carcinomas for expression of glial fibrillary acidic protein. *Am J Surg Pathol* 5:710–718
7. Ishihara S, Honda Y, Iyama K (2007) A case of extraneural ependymoma of the abdominal cavity. *Jpn J Diagn Pathol* 4:440–444
8. Deval B, Rousset P, Bigenwald C et al (2014) Treatment of ovarian anaplastic ependymoma by an aromatase inhibitor. *Obstet Gynecol* 123:488–491
9. Okazaki H (1997) A case of ependymoma ovarii. *Acta Obst Gynaec Jpn* 49:815–817
10. Mikami M, Yoshii T, Sakaiya N et al (2000) A case of ependymoma of ovary that long term oral administration of VP-16 was effective for prolongation for disease-free interval. *Saitama Dist J Jpn Soc Obstet Gynecol* 30:25–29 (**Article in Japanese**)
11. Matsuyama A, Hisaoka M, Yamamoto I et al (2010) Extraspinal ependymoma of the broad ligament. *Pathol Int* 60:241–244
12. Ishikura H (2006) Ovary. *Pathol Clin Med* 24:240–246