

Primary retroperitoneal mature cystic teratoma with focal enteric type adenocarcinoma in a post-partum woman: report of a case with literature review

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Abstract

Teratomas are characterized by containing tissue from all three germinal cell layers. Occasionally, somatic type malignancies develop within a mature cystic teratoma. We reported here a rare case of enteric type adenocarcinoma, with associated dysplastic epithelial precursor lesion, arising within a mature cystic teratoma in the retroperitoneum of a 30-year-old woman status post vaginal delivery 11 weeks earlier. The mass is 17.5 cm and cystic. A polypoid mass component measuring 4.7×4.2×2.5 cm was located inside the cystic component. Microscopically, the majority of the specimen was a mature cystic teratoma with all three germinal cell layers. The polypoid mass component was an adenocarcinoma with an adjacent dysplastic epithelial precursor lesion. The adenocarcinoma was diffusely positive for CK20 and CDX-2, and focally positive for CD7, indicating enteric differentiation. A brief review of retroperitoneal mature cystic teratomas with associated somatic type malignancy was performed.

Introduction

Teratomas are characterized by the presence of tissue arising from all three germinal layers and can occur in both gonadal and extragonadal sites along the midline including the mediastinum and retroperitoneum. Primary retroperitoneal teratoma is rare, and is commonly found in children and young adults.¹ Primary retroperitoneal teratomas can occasionally undergo malignant transformation. To our knowledge, only 10 cases have been reported in the English literature.²⁻¹¹

Herein, we report an enteric type adenocarcinoma arising in a primary retroperitoneal mature cystic teratoma (MCT) in a 30-year-old post-partum woman.

Case Report

The patient was a 30-year-old woman who gave birth to a full-term daughter by vaginal delivery on September 12, 2010. Three weeks later, she noticed a soft, palpable lump underneath her left ribs. Magnetic resonance imaging (MRI) of the abdomen and pelvis was performed and demonstrated a large, well-circumscribed heterogeneous signal intensity mass, measuring 13.0×11.3×14.0 cm centered in the left retroperitoneum (Figure 1A-D). Its dominant component was T1 hyperintense and T2 hypointense, suggesting proteinaceous fluid, although there were also areas of macroscopic fat content, along with several mural polypoid nodular enhancing components. This mass did not appear to arise directly from the left kidney or other adjacent parenchymal organs. The left kidney was displaced anteromedially, the left renal vein and splenic veins were displaced anterosuperiorly, and multiple small bowel loops were displaced to the right by the mass without gross invasion. The mass was also located adjacent to the distal transverse colon, left adrenal gland, and pancreatic tail without gross invasion. The uterus and ovaries appeared normal, and no lymphadenopathy was seen. Resection of the retroperitoneal mass with en block left nephrectomy was subsequently performed without subsequent administration of chemotherapy. The patient did well status post surgery for 7 months without evidence of metastasis.

Pathology

Macroscopically, the resected retroperitoneal mass including the left kidney measured 22.5×19.0×10.0 cm, where the mass alone measured 17.5×15.0×10.0 cm and the kidney alone measured 7.5×7.5×4.5 cm. The mass was serially sectioned to reveal a multicystic cut surface with green-brown fluid (~500 mL). Cheesy white-tan material was also noted. Further sectioning revealed a yellow-tan cut surface with calcified foci, consistent with teeth. In addition, a polypoid mass component was identified that measured 4.7×4.2×2.5 cm and protruded into the cystic component. The mass did not grossly invade the kidney.

Microscopically, a mature cystic teratoma with tissue arising from all of three germ cell layers, including squamous epithelium (Figure 2A), cartilage and adipose tissue (Figure 2B), and respiratory type mucosa (Figure 2C) was seen. No immature elements were seen. The grossly identified polypoid mass component

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was composed of moderately differentiated adenocarcinoma with infiltration into the cystic wall (Figure 2D-H), which did not extend beyond the cystic component wall. Benign glandular mucosa with an adjacent dysplastic epithelial precursor lesion was identified (Figure 2E). The adenocarcinoma was composed of glands with blue mucin in the lumens (Figure 2D and 2F). The adenocarcinomatous glands consist of columnar cells with prominent nuclei and coarse chromatin. Tumor necrosis was common (Figure 2F and 2G). Brisk mitosis with atypical mitosis was identified (Figure 2H). The Immunohistochemical staining with adequate controls was performed to further characterize the lesion. The adenocarcinoma was diffusely positive for CK20 and CDX-2, and focally positive for CD7, indicating enteric differentiation.

Discussion

Extragenital teratomas are thought to arise from primordial germ cells or early embryonic cells,^{12,13} and arise from totipotential cells with a diploid chromosome.¹⁴ Primary retroperitoneal teratoma is rare, accounting for 1-11% of retroperitoneal neoplasms, and more frequently occur in neonates and young adults with a bimodal peak occurring in the first 6 months of life and again in young adults.¹⁵⁻¹⁸ Grossly, two variants, cystic or solid, are identified. Cystic teratomas consist of fully mature elements and are usually benign whereas the solid variant is more likely malignant.¹⁹ However, a quarter of benign teratomas may be

solid.²⁰ Microscopically, the cystic components in mature cystic teratomas are lined by different epithelial cells including squamous, ciliated, or glandular-like cells, and are composed of all types of mature tissue including epidermis, brain, glial tissue, teeth, cartilage, nerve, smooth muscle, respiratory and intestinal

epithelium. Presence of undifferentiated tissue, mostly neuroepithelial in origin, indicates that immature elements are present.¹³ It has been proposed to stratify teratomas based on the nature, proportion, and characteristics of tissues present within the tumor on condition of adequate sampling (at least 1 section per

centimeter of the largest dimension of the tumor and at least 10 sections).¹²

Approximately 1% of mature teratomas undergo malignant transformation.²¹ The prognosis for malignancy arising in the mature teratomas is largely derived from studies of ovarian mature cystic teratomas with malignant

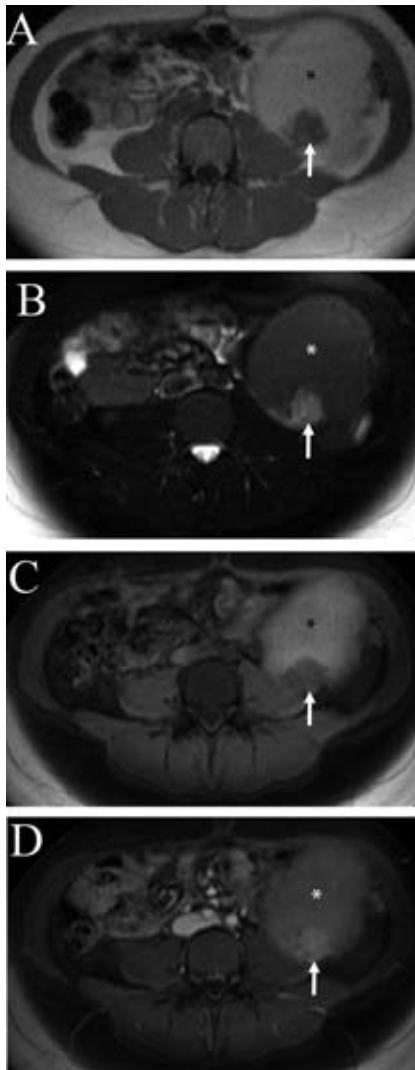


Figure 1. Abdominal magnetic resonance images of patient with left retroperitoneal mass. Axial T1-weighted gradient-echo images (A), axial fat-suppressed T2-weighted fast spin echo image (B), axial fat-suppressed T1-weighted gradient-echo image (C), and venous phase post-contrast axial fat-suppressed T1-weighted gradient echo image (D) show inferior portion of large retroperitoneal mass with dominant non-enhancing cystic component containing proteinaceous fluid (*) with high T1-weighted and intermediate-slightly high T2-weighted signal intensity relative to skeletal muscle, and mural polypoid nodular enhancing component posteriorly (arrow) with intermediate T1-weighted and high T2-weighted signal intensity relative to skeletal muscle.

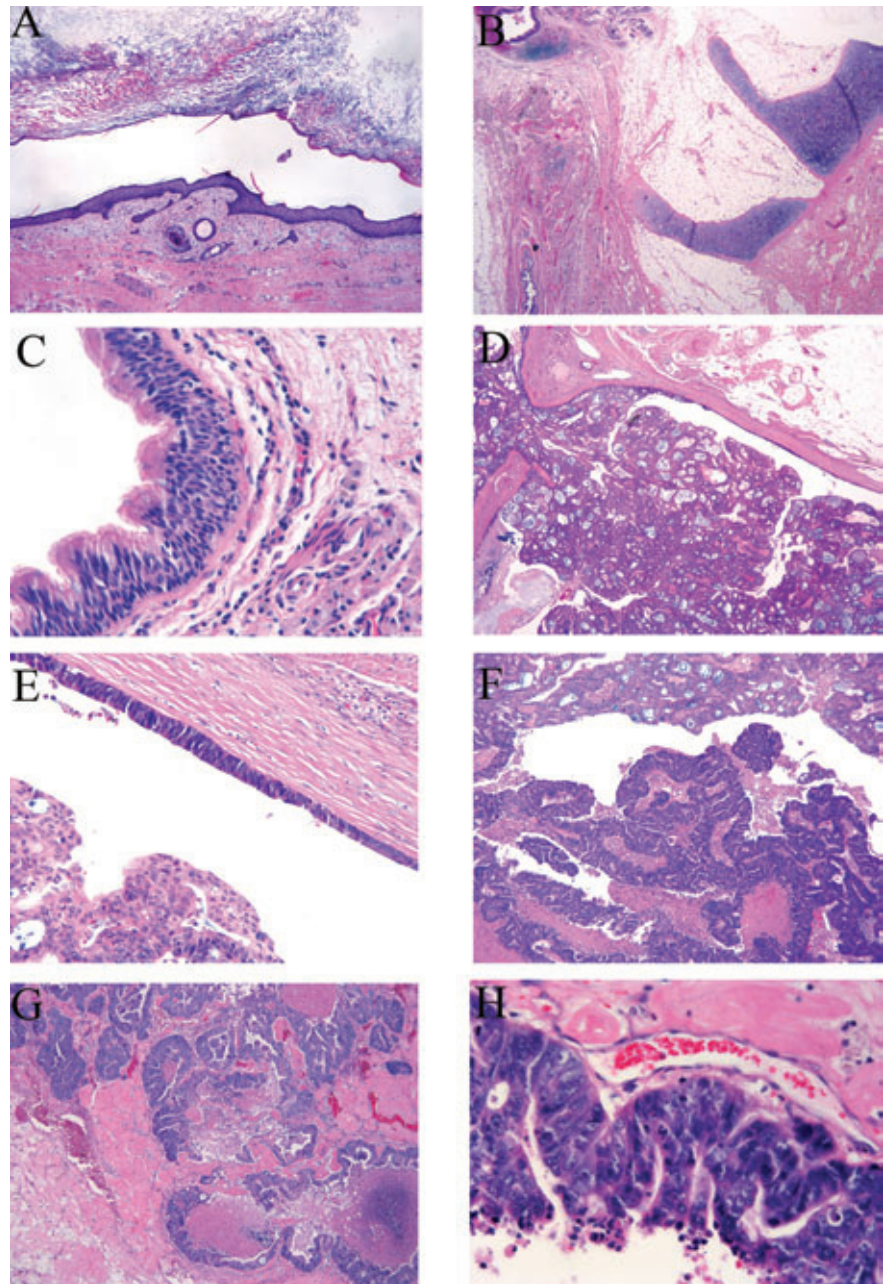


Figure 2. Mature cystic teratoma with an enteric type adenocarcinoma. Squamous epithelium with hair follicle, Haematoxylin and Eosin (H&E), x50 (A); cartilage and adipose tissue H&E, x25 (B); respiratory type epithelium, H&E, x400 (C); adenocarcinoma protruding into cystic component, H&E, x25 (D); adenocarcinoma and adjacent precursor lesion, H&E, x200 (E); adenocarcinoma with blue mucin and tumor necrosis, H&E, x50 (F); adenocarcinoma with cystic wall invasion, H&E, x50 (G); adenocarcinoma with brisk mitosis including atypical mitosis.

Table 1. Summary of reported mature cystic teratomas with malignant transformation in the literature.

Authors	Sex/age	Presenting symptoms	Size (cm)	Somatic type malignancy type	Somatic type malignancy size (cm)	Follow-up
Wang <i>et al.</i> ¹⁰	M/40	Right back pain	9	Adenocarcinoma	NS	DOD 16 M
Yamasaki <i>et al.</i> ¹¹	F/53	Urinary frequency	12×6×6	Carcinoid tumor	NS	ANED 31 M
Song <i>et al.</i> ²³	F/73	Palpable mass for 10 M	12×12×10	Mucinous adenocarcinoma	1.5×1	DOD 2 M
Chang <i>et al.</i> ²	M/35	Low back pain for 5 yr	20×8×8	Gastrointestinal type adenocarcinoma	2.4×1.4×0.6	ANED 2 yr
Chu <i>et al.</i> ⁴	F/35	Left abdominal pain, weight loss	14×10×9	Signet ring and intestinal type adenocarcinoma	NS	DOD 2 M
Joseph <i>et al.</i> ⁵	F/67	Left abdominal pain, weight loss, constipation for 6 M	10×6×3	Squamous cell carcinoma	3×1.3	NS
Marusic <i>et al.</i> ⁶	M/36	Dyspnea	16	Papillary renal cell like carcinoma	NS	ANED 6 M
Cheung <i>et al.</i> ³	F/47	Right abdominal pain and distension for 2 M, weight gain of 2 kg	20×18×10	Colonic type adenocarcinoma	1.5	ANED 18 M
Scott <i>et al.</i> ⁷	F/48	Left upper quadrant pain	15.2×11.6×14.1	Neuroendocrine carcinoma	1.2	ANED 6 M
Torado <i>et al.</i> ⁸	M/28	Abdominal pain	10×7	Small intestinal type adenocarcinoma in situ	NS	DOD 10 M
Present case	F/30	Palpable mass	17.5×15.0×10.0	Enteric type adenocarcinoma	4.7×4.2×2.5	ANED 7 M

ANED, alive without evidence of disease; DOD, dead of disease; NS, not stated; M, month.

transformation, which show a poor prognosis. Factors adversely affecting patient prognosis include tumor extension beyond the cyst wall, cystic wall disruption, higher tumor grade, and histological type.²² Whether such criteria can be applied to retroperitoneal teratomas with malignant transformation is still undetermined. In this report, we also summarize 11 cases of mature teratomas with malignant transformation from the literature (Table 1),^{2-8,10,11,23} of which 7 cases were adenocarcinoma (5 enteric type, 1 mucinous, 1 not otherwise specified). In addition, there is one case each for neuroendocrine carcinoma, carcinoid, squamous cell carcinoma, and papillary renal cell-like carcinoma. Five patients presented with abdominal pain, two with back pain, two with a palpable mass lesion, one with dyspnea, and one with urinary frequency. The tumors are usually large, ranging from 9 to 20 cm in size, with an average greatest dimension of 14.2 cm. The number of retroperitoneal mature cystic teratomas with associated carcinoma is too few and makes the evaluation of their prognosis unreliable. These tumors may show a similar or worse prognosis compared with those arising from the ovaries, considering their anatomic location and frequent delayed clinical diagnosis. Evaluation for local invasion, positive surgical margins, and presence of metastases is warranted for these tumors. The treatment for these tumors is typically complete resection, which is critical for both diagnosis and management. Little data are available to evaluate the benefit of chemotherapy and radiation in this clinical scenario, although studies of ovarian teratomas with malignant transformation have shown that they are generally resistant to chemotherapy and radiation therapy.²⁴

In summary, we describe a case of mature

cystic teratoma with malignant transformation in a 30-year-old postpartum woman. A mature cystic teratoma was recognized with all three germinal cell layers. An enteric type adenocarcinoma with an adjacent dysplastic epithelial precursor lesion was identified. Immunohistochemical staining showed that the carcinoma was floridly positive for CK20 and CDX2 and focally positive for CK7. These findings supported an enteric differentiation of adenocarcinoma arising from the mature cystic teratoma.

References

- Barksdale EM Jr, Obokhare I. Teratomas in infants and children. *Curr Opin Pediatr* 2009;21:344-9.
- Chang YL, Wu CT, Lee YC. Mediastinal and retroperitoneal teratoma with focal gastrointestinal adenocarcinoma. *J Thorac Oncol* 2006;1:729-31.
- Cheung WL, Cao D. Colonic-type adenocarcinoma arising in a primary retroperitoneal mature cystic teratoma. *Pathol Int* 2008;58:792-6.
- Chu PY, Teng TH, Lee CC, Chou YY. Adenocarcinomas arising from primary retroperitoneal teratoma in an adult female patient. *Int J Urol* 2006;13:1352-4.
- Joseph LD, Devi MK, Sundaram S, Rajendiran S. Squamous cell carcinoma arising from primary retroperitoneal mature teratoma. *J Assoc Physicians India* 2007;55:374-6.
- Marusic Z, Zovak M, Hagenkord JM, et al. Papillary renal cell-like carcinoma in a retroperitoneal teratoma. *Pathol Int* 2010;60:581-5.
- Scott AL, Abbassi-Ghadi N, Archer CM, et al. Neuroendocrine carcinoma arising within a retroperitoneal mature teratoma. *Ann R Coll Surg Engl* 2010;92:W5-8.
- Terado Y, Kurata A, Ishida T, et al. Adenocarcinoma of small intestinal type in retroperitoneal mature teratoma. *Pathol Int* 2010;60:701-5.
- Tezel E, Sare M, Edali N, et al. Retroperitoneal malignant teratoma. A case report. *Mater Med Pol* 1995;27:123-5.
- Wang LJ, Chu SH, Ng KF, Wong YC. Adenocarcinomas arising from primary retroperitoneal mature teratomas: CT and MR imaging. *Eur Radiol* 2002;12:1546-9.
- Yamasaki T, Yagihashi Y, Shirahase T, et al. Primary carcinoid tumor arising in a retroperitoneal mature teratoma in an adult. *Int J Urol* 2004;11:912-5.
- Gatcombe HG, Assikis V, Kooby D, Johnstone PA. Primary retroperitoneal teratomas: a review of the literature. *J Surg Oncol* 2004;86:107-13.
- Saiga T, Osasa H, Hatayama H, et al. The origin of extragonadal teratoma: case report of an immature teratoma occurring in a prenatal brain. *Pediatr Pathol* 1991;11:759-70.
- Wagner H, Baretton GB, Schneiderbanger K, et al. Sex chromosome determination in extragonadal teratomas by interphase cytogenetics: clues to histogenesis. *Pediatr Pathol Lab Med* 1997;17:401-12.
- Gschwend J, Burke TW, Woodward JE, Heller PB. Retroperitoneal teratoma presenting as an abdominal-pelvic mass. *Obstet Gynecol* 1987;70:500-2.
- Lane RH, Stephens DH, Reiman HM. Primary retroperitoneal neoplasms: CT findings in 90 cases with clinical and pathologic correlation. *AJR Am J Roentgenol* 1989;152:83-9.
- Panageas E. General diagnosis case of the

- day. Primary retroperitoneal teratoma. *AJR Am J Roentgenol* 1991;156:1292-4.
18. Wang RM, Chen CA. Primary retroperitoneal teratoma. *Acta Obstet Gynecol Scand* 2000;79:707-8.
19. Bellin MF, Duron JJ, Curet P, et al. Primary retroperitoneal teratoma in the adult: correlation of MRI features with CT and pathology. *Magn Reson Imaging*. 1991;9:263-6.
20. Davidson AJ, Hartman DS, Goldman SM. Mature teratoma of the retroperitoneum: radiologic, pathologic, and clinical correlation. *Radiology* 1989;172:421-5.
21. Renato F, Paolo V, Girolamo M, et al. Malignant retroperitoneal teratoma: case report and literature review. *Acta Urol Belg* 1996;64:49-54.
22. Templeman CL, Fallat ME, Lam AM, et al. Managing mature cystic teratomas of the ovary. *Obstet Gynecol Surv* 2000;55:738-45.
23. Song ES, Choi SJ, Kim L, et al. Mucinous adenocarcinoma arising from one retroperitoneal mature cystic teratoma in a postmenopausal woman. *J Obstet Gynaecol Res* 2005;31:127-32.
24. Donadio AC, Motzer RJ, Bajorin DF, et al. Chemotherapy for teratoma with malignant transformation. *J Clin Oncol* 2003; 21:4285-91.