

Abdominal wall mixed malignant germ cell tumor: a case report and review of literature

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Summary

Abdominal wall mass is not uncommon in clinic, but it is very rare that germ cell tumors (GCTs) arise in the abdominal wall. The authors review the case of a 34-year-old female with abdominal wall mixed malignant GCT composed of embryonal carcinoma and teratoma and combine the relative literature to explain why GCTs originate from anterior abdominal wall.

Key words: Germ cell tumors; Extragenital; Abdominal wall.

Introduction

Germ cell tumors (GCTs) most frequently arise in the gonads, but extragenital germ cell tumors (EGCTs) are estimated to represent only 2-5% of all adult GCTs [1]. The most common primary site of EGCTs is midline structures such as the anterior mediastinum and retroperitoneum, but also can be described in sacrococcygeal region, rectum, head and neck soft tissues, nasal cavity, and paranasal sinuses [2-4]. Primary sites in abdominal wall have been found in only two reports, one is malignant teratoma [5] and another is yolk sac tumor [6]. To the best of the present authors' knowledge, the reported case in this article of abdominal wall mixed malignant germ cell tumor consisting of embryonal carcinoma and teratoma components has not been reported previously.

Case Report

A 34-year-old female with a large abdominal mass which could be touched at six months before admission was treated in The First Affiliated Hospital of Guangxi Medical University on July 16, 2014. The patient had suffered from a dull aching pain in the left abdomen for 15 days and had worsened in the last two days. Physical examination showed a 90×100×80 mm mass with well margin was located at the left adnexa which caused tenderness, was firm, and freely movable.

Two-dimensional pelvic ultrasound examination showed a 97×108×93 mm mixed echoic mass in the left hypogastric region. Laboratory data showed elevated serum level of alpha-fetoprotein (AFP; 10,382 ng/ml, normal range 0-11.00 ng/ml), cancer antigen 125 (CA125; 124.00 U/ml, normal range 0-35.00 U/ml), and cancer antigen 19-9 (CA19-9; 52.98 U/ml, normal range 10.00-37.00 U/ml). The patient received exploratory laparotomy after her consent was obtained. A mass of 110×100×90 mm located in preperitoneal space between rectus abdominis was found during surgery. A simple total tyelectomy

was performed. Histopathologic features were consistent with abdominal wall mixed malignant germ cell tumor composed of embryonal carcinoma (Figures 1a, 1b) and teratoma (Figure 1c). In immunohistochemical assay, the tumor tissue staining was positive for placental alkaline phosphatase (PLAP) (Figure 1d) and cellular keratin (CK) (Figure 1e), but negative for AFP (Figure 1f), CD30, and β -human chorionic gonadotropin (hCG).

Five days after the total tyelectomy, the serum level of AFP and CA125 decreased to 2,552 ng/ml and 83.93 U/ml, respectively. The level of CA19-9 was in the normal range. The scheduled chemotherapy of bleomycin, etoposide, and cisplatin (BEP) regimen was given to the patient postoperatively. The patient refused to accept further chemotherapy due to insufficient funds after two cycles of BEP regimen. The patient was well with normal menstrual cycle and there was no evidence of tumor recurrence in the eight-month's follow-up.

Discussion

The tissue source of EGCTs is not clear at present. Most people believe that a mismigration of primordial germ cells (PGCs) along the urogenital ridge during yolk sac as a result of the influence of some factors in early embryogenesis [7, 8]. They also may result from germ cells that are distributed physiologically to other tissues or organs to provide regular functions or convey hematologic or immunologic information [9]. In this patient, a mass with the pathologic diagnosis of mixed malignant GCTs comprised of embryonal carcinoma and teratoma components in the anterior abdominal wall was found. Could GCTs originate from anterior abdominal wall? We know that PGCs arise in the endoderm of the embryonic yolk sac which is wrapped around the umbilical cord. The germ cells migrate through the wall of the midgut to the genital ridge at four to five weeks of gestation [10]. In the process of migration, PGCs can be lost in the abdominal

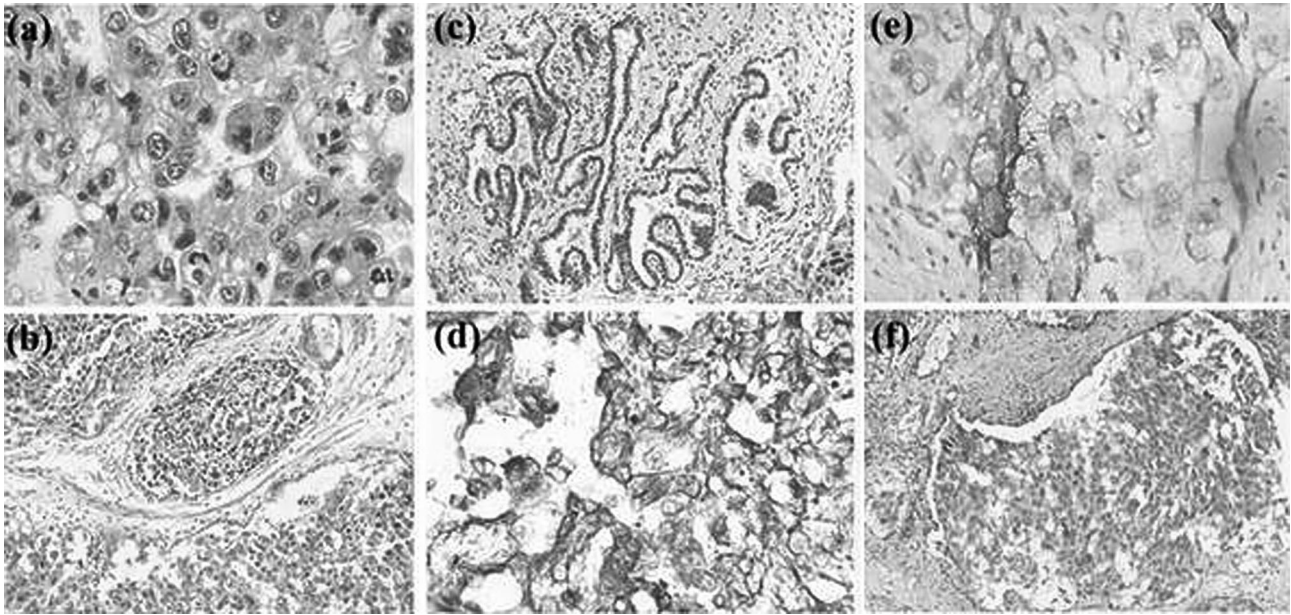


Figure 1. — (a) Embryonal carcinoma. Pleomorphic cells and fission are easily seen (HE $\times 400$). (b) Embryonal carcinoma. Tumor cells infiltrating the muscle layer of abdominal (HE $\times 100$). (c) Teratoma. Photomicrograph showing differentiated gland structures (HE $\times 100$). (d) Immunohistochemistry for PLAP showing strong cytoplasmic positivity in most of the large tumor cells. (e) Immunohistochemistry for CK showing positivity in some tumor cells. (f) Immunohistochemistry for AFP showing negativity in all tumor cells.

wall under some circumstances, and then the tumor will occur. In this patient, the authors speculate that the tumor located between preperitoneal space and rectus abdominis initiate from some aberrant cells of mesoderm near the yolk sac based on the mechanism mentioned above.

EGCTs lack specificity in clinical manifestation, therefore preoperative diagnosis is difficult. Most cases were diagnosed after surgical exploration, and the confirmed diagnosis is still based on pathological result. Some immunohistochemical markers can be used in differential diagnosis. PLAP is considered to be expressed specifically in GCTs [11-16]. AFP is a characteristic marker in yolk sac tumors. It can also be expressed in a handful of embryonal carcinoma and immature teratoma. The majority of choriocarcinoma show immunopositivity for hCG, and CK is positive in embryonal carcinoma, choriocarcinoma, immature teratoma, and yolk sac tumors [15]. CD30 has been found highly expression in embryonal carcinoma, but not every case of embryonal carcinoma shows positive results for CD30 [16, 17]. In this case, PLAP was positive stained in tumor tissue which was highly suggestive the diagnosis of GCTs. The positive staining of CK, negative staining of AFP, HCG, and CD30 support the tumor's embryonic carcinomas and teratoma immunophenotype and exclude the diagnosis of yolk sac tumor.

EGCTs is highly malignant, however with a promising

prognosis. Surgery is the first line of choice for this type of disease with the option of chemotherapy as adjunctive treatment. Chemotherapeutic regimen of BEP used for gonadal GCTs, especially those arising in the testis, also has been demonstrated effective in EGCTs. Platinum-based chemotherapy following complete resection of the tumor may significantly improve the prognosis of the disease. However the survival rate of EGCTs is different in the related reports [18-20]. In the present case, the patient's prognosis was favorable so far although she just received incomplete chemotherapy after operation.

Conclusion

The present case report has highlighted that a mass in the abdominal wall should not be neglected as extragonadal germ cell tumor. The histopathologic examination is the gold standard for diagnosis. The authors hope that these experiences may provide some help for clinicians.

References

- [1] Stang A., Trabert B., Wentzensen N., Cook M.B., Rusner C., Oosterhuis J.W., MoGlynn K.A.: "Gonadal and extragonadal germ cell tumours in the United States, 1973-2007". *Int. J. Androl.*, 2012, 35, 616.
- [2] Miller K.D., Michael H., Jacobson L., Sutton G.P.: "Primary yolk sac tumor of the rectum". *Cancer Invest.*, 2000, 18, 597.
- [3] Dehner L.P., Mills A., Talerman A., Billman G.F., Krous H.F., Platz

- C.E.: "Germ cell neoplasms of head and neck soft tissues: a pathological spectrum of teratomatous and endodermal sinus tumors". *Hum. Pathol.*, 1990, 21, 309.
- [4] Heffner D.K., Hyams V.J.: "Teratocarcinoma (malignant teratoma?) of the nasal cavity and paranasal sinuses. A clinicopathologic study of 20 cases". *Cancer*, 1984, 53, 2140.
- [5] Rao K., Dutta T.K., Gupta A.N., Aikat M.: "Extragenadal abdominal wall malignant teratoma--a case report". *Indian J. Cancer*, 1980, 17, 185.
- [6] Jacob R., Ramadas K., Jyothirmayi R., Kusumakumary P., Nair M.K.: "Extragenadal germ-cell tumors: a ten-year experience". *Am. J. Clin. Oncol.*, 1998, 21, 198.
- [7] Bokemeyer C., Nichols C.R., Droz J.P., Schmoll H.J., Horwich A., Gerl A., et al.: "Extragenadal germ cell tumors of the mediastinum and retroperitoneum: results from an international analysis". *J. Clin. Oncol.*, 2002, 20, 1864.
- [8] Rusner C., Trabert B., Katalinic A., Kieschke J., Emrich K., Stang A.: "Incidence patterns and trends of malignant gonadal and extragonadal germ cell tumors in Germany, 1998-2008". *Cancer Epidemiol.*, 2013, 37, 370.
- [9] Friedman N.B.: "The function of the primordial germ cell in extragonadal tissues". *Int. J. Androl.*, 1987, 10, 43.
- [10] Legato M.J.: "Principles of gender-specific medicine". In: Legato M.J.(ed). *Difference in germ cell tumors of the reproductive tract in men and women*. 2nd ed. New York: Academic Press, 2009, 12.
- [11] Hattab E.M., Tu P.H., Wilson J.D., Cheng L.: "OCT4 immunohistochemistry is superior to placental alkaline phosphatase (PLAP) in the diagnosis of central nervous system germinoma". *Am. J. Surg. Pathol.*, 2005, 29, 368.
- [12] Stoop H., Honecker F., van de Geijn G.J., Gillis A.J., Cools M.C., de Boer M., et al.: "Stem cell factor as a novel diagnostic marker for early malignant germ cells". *J. Pathol.*, 2008, 216, 43.
- [13] Liu A., Cheng L., Du J., Peng Y., Allan R.W., Wei L., et al.: "Diagnostic utility of novel stem cell markers SALL4, OCT4, NANOG, SOX2, UTF1, and TCL1 in primary mediastinal germ cell tumors". *Am. J. Surg. Pathol.*, 2010, 34, 697.
- [14] Mueller T., Mueller L.P., Holzhausen H.J., Witthuhn R., Albers P., Schmoll H.J.: "Histological evidence for the existence of germ cell tumor cells showing embryonal carcinoma morphology but lacking OCT4 expression and cisplatin sensitivity". *Histochem. Cell. Biol.*, 2010, 134, 197.
- [15] Sakurada K., Saino M., Mouri W., Sato A., Kitanaka C., Kayama T.: "Nestin expression in central nervous system germ cell tumors". *Neurosurg. Rev.*, 2008, 31, 173.
- [16] Gao Y., Jiang J., Liu Q.: "Clinicopathological and immunohistochemical features of primary central nervous system germ cell tumors: a 24-years experience". *Int. J. Clin. Exp. Pathol.*, 2014, 7, 6965.
- [17] Deutsch Y.E., Tadmor T., Podack E.R., Rosenblatt J.D.: "CD30: an important new target in hematologic malignancies". *Leuk. Lymphoma*, 2011, 52, 1641.
- [18] Albany C., Einhorn L.H.: "Extragenadal germ cell tumors: clinical presentation and management". *Curr. Opin. Oncol.*, 2013, 25, 261.
- [19] Fukui N., Kohno Y., Ishioka J., Fukuda H., Kageyama Y., Higashi Y.: "Treatment outcome of patients with extragonadal nonseminomatous germ cell tumors: the Saitama Cancer Center experience". *Int. J. Clin. Oncol.*, 2013, 18, 731.
- [20] Rodney A.J., Tannir N.M., Siefker-Radtke A.O., Liu P., Walsh G.L., Milliken R.E., et al.: "Survival outcomes for men with mediastinal germ-cell tumors: the University of Texas M. D. Anderson Cancer Center experience". *Urol. Oncol.*, 2012, 30, 879.

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