Intra-abdominal desmoplastic small round cell tumor in a 68-year-old female

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Summary

Background: Desmoplastic small round cell tumor (DSRCT) of the peritoneum typically occurs in young adults. The mean age of females with DSRCT is 20 years. We describe a DSRCT with an unusual age of presentation mimicking a metastatic ovarian neoplasm.

Case: A 68-year-old para 4 female presented with abdominal enlargement. Laparatomy showed multiple tumor nodules attached to the peritoneal surface. The tumor was debulked. The histological findings were characteristic for DSRCT. Adjuvantly the patient received cytotoxic chemotherapy but died of recurrent disease 3 months after initial diagnosis.

Conclusion: DSRCT should be added to the differential diagnosis of unusual gynecologic malignancies in elderly as well as younger females. Identification of DSRCT is important because it can be confused with primary ovarian neoplasms.

Key words: Peritoneal neoplasm; Desmoplastic small round cell tumor; Ovarian tumor.

Background

Many neoplasms that are specific to the female genital tract can involve its components as primary or secondary tumors. Accurate identification of such tumors is important because they can mimic primary neoplasms with substantially different biological behaviors, treatments and prognosis. We describe an intra-abdominal desmoplastic small round cell tumor (DSRCT) with an unusual age of presentation mimicking a metastatic ovarian neoplasm.

Case

A 68-year-old para 4 female presented with abdominal pain and enlargement. Clinical examination showed a distended abdomen with ascites and nodularity in the cul-de-sac highly suspicious for a metastatic ovarian neoplasm. Laboratory findings were unspecific. The serum CA 125 level was 79.1 IU/mL and calcium was 2.11 mmol/L. Computed tomography (CT) showed pelvic and abdominal tumor masses and ascites. Laparatomy revealed 3.5 L ascites and multiple grayish-white firm tumor nodules up to 18 cm in diameter attached to the peritoneal surface of the pelvis and abdomen. Multiple smaller satellite nodules, some of them pedunculated, were seen. The visceral peritoneum of the small and large bowel and liver were involved, as was the omentum. Some of the nodules were necrotic and hemorrhagic. The retroperitoneal lymph nodes were not palpably enlarged and there was no evidence of parenchymal organ involvement. The uterus, fallopian tubes and ovaries were free of disease. The peritoneal tumor was debulked with residual disease and hysterectomy with bilateral salpingo-oophorectomy and omentectomy was performed. The histologic and immunohistochemical findings were characteristic of DSRCT [1]. Adjuvantly the patient received vincristin (1.5 mg/m^2), methotrexate (7 mg/m^2) and cyclophosphamide (600 mg/m^2) but died of recurrent disease 3 months after the initial diagnosis.

Comment

DSRCT is a rare condition that usually diffusely involves the abdominal and/or pelvic peritoneum and pursues an aggressive clinical course [1]. It has been suggested that these tumors may be blastomas arising from intraembryonic mesoderm or coelom. Histology shows a nesting growth pattern of small basophilic tumor cells, prominant desmoplasia and multi-immunophenotypic differentiation with coexpression of epithelial, mesenchymal, and neuronal markers [1]. Approximately 100 cases have been reported mostly in male children and young adults. Only 21 cases have been described in females [2]. Ovarian involvement was found in seven and uterine involvement in two of these 21 cases [2]. The mean age of females with DSRCT is 20.4 (5-63) years [2] and only three cases have been reported in postmenopausal females [2-4]. Mead *et al.* [3] described a DSRCT in a 52-year-old woman with signs and symptoms that mimicked ovarian carcinoma. A 60-year-old patient presenting with an acute abdomen was reported by Fukunaga *et al.* [4]. Hui *et al.* [2] described a 63-year-old female with DSRCT presenting with hematuria. Our patient is the oldest reported female with DSRCT.

This case suggests that DSRCT should be added to the differential diagnosis of unusual gynecologic malignancies in elderly as well as younger females.

Identification of DSRCT is important because it can be confused with primary ovarian neoplasms clinically, by imaging studies or at laparotomy. Treatment consists of palliative tumor removal. No effective chemotherapy or radiotherapy regimes have been reported.

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