

CASE REPORT

Endodermal sinus tumor: a rare cause of calcified peritoneal implants

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Abstract

Calcified peritoneal implants have been attributed to various malignant and benign causes. We present an interesting case of a 32-year-old woman who presented with abdominal pain, distension and vaginal bleeding. Contrast-enhanced computed tomography revealed calcified peritoneal implants with a malignant ovarian mass. Histopathology showed an endodermal sinus tumor, a rare cause of calcified peritoneal carcinomatosis.

Keywords: Endodermal sinus tumor; ovary; yolk sac tumor; computed tomography.

Metastatic calcified peritoneal implants are rarely encountered in abdominopelvic computed tomography (CT). Ovarian carcinoma is the most common malignant cause of calcified peritoneal carcinomatosis. Serous cystadenocarcinoma produces radiographically visible calcification in 12% of cases.^[1] Calcification in peritoneal metastasis of endodermal sinus tumor is not described in literature. We report a case of endodermal sinus tumor of the ovary with calcified peritoneal implants at the time of presentation.

Case

A 32-year-old woman presented with abdominal pain and distension for 2 months and vaginal bleeding of 1 month duration. She had fever off and on. There was loss of appetite and loss of weight. On palpation, a vague mass was felt in the lower abdomen. Per vaginal examination revealed a normal anteverted uterus. A 5×6 cm size mass was felt in the left fornix, which was firm, mobile and non-tender. A markedly elevated serum alpha-fetoprotein (AFP) level to 11.357 ng/ml suggested the diagnosis of an ovarian yolk sac tumor. Serum human gonadotropin hormone levels and CA-125 levels were within normal limits. All other biochemical and laboratory

investigations, including serum urea and creatinine, were also normal. The patient had no significant past or family history. Subsequently, contrast-enhanced CT of the abdomen was performed, which revealed a 4.5×5.2×6 cm size complex mass lesion in the left adnexa. A well-defined enhancing solid component was seen with cystic areas adjacent to it. Few hyperdense calcific specks were present within the solid component. The uterine body was displaced to the contralateral side by the ovarian mass. Enhancing soft tissue density nodular lesions were seen in the peritoneal reflections along the left paracolic gutter and pelvis (Fig. 1a,b). Coarsened nodular and curvilinear sheetlike hyperdensities (attenuation approaching that of bone) were present, distributed along the undersurface of the hemidiaphragm, the perihepatic region and Morrison's pouch (Fig. 2). Mild ascites was also present in the abdomen and cul-de-sac. These findings were suggestive of yolk sac tumor of the left ovary (in view of the raised AFP level) with calcified peritoneal carcinomatosis. Fine-needle aspiration cytology was obtained, which further confirmed the presumptive diagnosis of endodermal sinus tumor. The smear showed tumor cells arranged in papillary groups; tight cell clusters were seen forming a glandular patterned acinar structure with a central

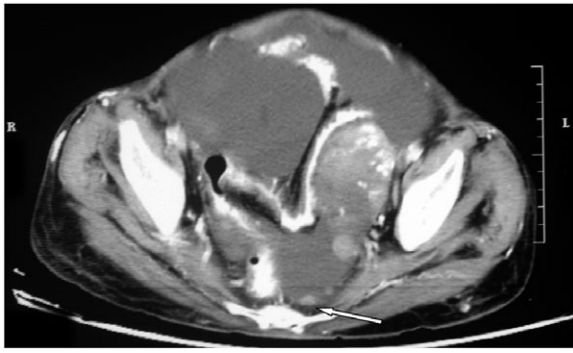


Figure 1 Contrast-enhanced axial CT section through the pelvis: a complex left ovarian mass with pelvic peritoneal metastasis (arrows).

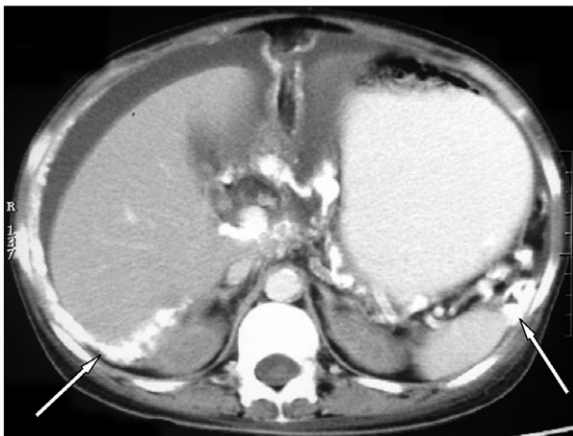
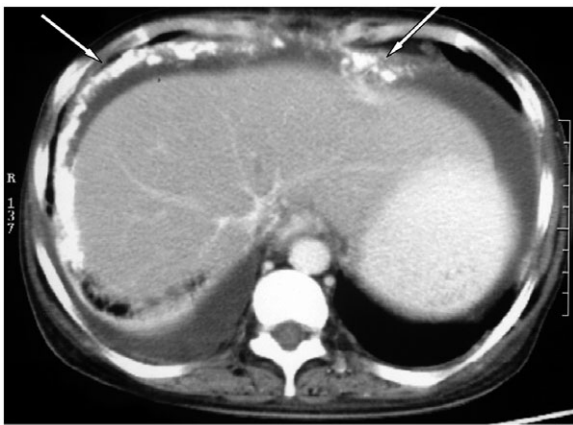


Figure 2 Contrast-enhanced CT of the abdomen showing calcified peritoneal implants in the undersurface of the right dome of the diaphragm, perihepatic and perisplenic region, and Morrison's pouch (arrows).

capillary (Schiller–Duval body). Enlarged hyperchromatic nuclei and a moderate amount of cytoplasm were present. However, a histological diagnosis could not be ascertained as the patient did not undergo surgery. Adjuvant combination chemotherapy was administered.

Discussion

Peritoneal carcinomatosis is the most common route of spread of ovarian malignancy. Almost 90% cases of carcinoma ovary show metastasis along the peritoneal surface at autopsy.^[2] Metastatic malignant peritoneal calcification is most frequently seen in serous cystadenocarcinoma, the most common type of ovarian malignancy, which also shows histological calcification in nearly 30% cases.^[3] The other malignancies that may cause peritoneal calcification are primary papillary serous peritoneal carcinoma,^[4] colon cancer,^[5] gastric cancer^[6] and also squamous cell lung cancer, renal cell carcinoma, and melanoma, which induce paraneoplastic hyperparathyroidism and hypercalcemia.^[7]

Deposition of calcium in peritoneal implants occurs by metastatic and dystrophic calcification. Systemic causes of mineral imbalance, such as uremia or hyperparathyroidism, cause metastatic calcification; local tissue injury, the aging process or disease including malignancy cause a dystrophic type of calcification.^[8]

Peritoneal calcification is classified based on its morphological features. Circumscribed or focal calcification is described as nodular, and flat curvilinear calcification extending along the peritoneal plane as sheetlike. Although sheetlike calcification is more commonly associated with benign causes of peritoneal calcification (peritoneal dialysis, tuberculosis) it may be seen in its malignant counterpart (22%).^[4] Calcified peritoneal metastasis has not been described in yolk sac tumor of the ovary before.

Endodermal sinus tumor of the ovary, also known as yolk sac tumor, is a rare complex malignant ovarian tumor of germ cell origin that occurs in girls and young women, usually in the second decade of life (mean age 19 years).^[9] All malignant germ cell tumors constitute about 5% and endodermal sinus tumor constitute <1% of total malignant ovarian neoplasms. Yolk sac tumor is the second most common germ cell tumor. It is unilateral in 99% of cases. The diameter of this aggressive tumor ranges from 7 to 28 cm, with a median of 15 cm. Yolk sac tumors exhibit malignant changes in a cell line committed to extra embryonic differentiation and secrete alpha-fetoprotein. The cut surface of the tumor shows cystic and solid areas along with large areas of hemorrhage and necrosis. Cysts of varying sizes are diffusely scattered throughout the tissue, giving the tumor a honeycombed appearance. Microscopically, the many endodermal sinus patterns may include reticular, papillary, solid, polyvesicular vitelline, and polyembryonal histology. Schiller–Duval bodies are characteristically also present.^[10] The presence of calcification within the tumor often indicates its origin from a teratodermoid.

Imaging features consist of a complex mass of mixed solid and cystic nature. The tumor may be predominantly solid with areas of hemorrhage and cysts, displaying a heterogeneous appearance. Ultrasonography reveals a

pelvic mass depicting both echogenic solid and hypo/anechoic cystic components. Ultrasonography may also show concurrent dermoid cyst of the ovary (occurring in 14% cases), ascites, or urinary obstruction. However, the cystic nature of the mass is more easily appreciated on CT, probably because of the hemorrhage or protein content and multiple internal septations.^[11] CT is also useful in further characterizing and staging the mass. As extensive areas of hemorrhage and necrosis are common and striking findings in endodermal sinus tumor, identification of hemorrhage on magnetic resonance imaging (MRI) as high intensity foci on T1-weighted images supports the diagnosis. Hypervascularity in the form of prominent enhancement on post-contrast CT images or signal voids on MRI may also be seen. Metastases to the peritoneum, bowel serosa, omentum, liver or lymph nodes occur in 30% of cases.^[10] However, calcified peritoneal implants seen in the index case are not documented in the literature.

The differential diagnoses of yolk sac tumor in a young female with a complex solid cystic abdominopelvic mass on imaging are cystic teratoma, tuboovarian abscess, appendiceal abscess, mesenteric cyst, or gastrointestinal duplication cyst.^[10] The serum alpha-fetoprotein level clinches the diagnosis. As endodermal sinus tumor exhibits rapid growth due its aggressive nature, a delay in diagnosis and subsequent treatment may harm the patient and sharply reduce the chances of survival, especially in patients with advanced disease.

With surgery alone, survival is rare for these radio-resistant tumors. Prior to the introduction of effective combination chemotherapy, the prognosis for patients with endodermal sinus tumor was poor; with a 3-year survival rate of 13%. However, now the survival rates have risen to 82.5% for patients receiving cisplatin, vinblastine, and bleomycin combination chemotherapy. A high percentage of cases of clinically aggressive malignant ovarian germ cell tumors respond well to cisplatin-based chemotherapy and are curable.^[13,14]

References

- [1] Mitchell DG, Hill MC, Hill S, Zaloudek C. Serous carcinoma of the ovary: CT identification of metastatic calcified implants. *Radiology* 1986; 158: 649–52.
- [2] Pannu HK, Bristow RE, Montz FJ, Fishman EK. Multidetector CT of peritoneal carcinomatosis from ovarian cancer. *Radiographics* 2003; 23: 687–701. doi:10.1148/rg.233025105. PMID:12740470.
- [3] Ferenczy A, Lalens M, Zoghby M, Hussain SS. Ultrastructural studies on the morphogenesis of psammoma bodies in ovarian serous neoplasia. *Cancer* 1977; 39: 2451–9. doi:10.1002/1097-0142(197706)39:6<2451::AID-CNCR2820390623>3.0.CO;2;7. PMID:872045.
- [4] Agarwal A, Yeh BM, Breiman RS, Qayyum A, Coakley FV. Peritoneal calcification: causes and distinguishing features on CT. *Am J Roentgenol* 2004; 182: 441–5.
- [5] Wong LL, Peh WC. Clinics in diagnostic imaging (22): calcified peritoneal carcinomatosis. *Singapore Med J* 1997; 38: 88–91.
- [6] Kunieda K, Okuhira M, Nakano T, *et al.* Diffuse calcification in gastric cancer. *J Int Med Res* 1990; 18: 506–14.
- [7] Grill V, Martin TJ. Hypercalcemia of malignancy. *Rev Endocr Metab Disord* 2000; 1: 253–63. doi:10.1023/A:1026597816193. PMID:11706739.
- [8] Giachelli CM. Ectopic calcification: gathering hard facts about soft tissue mineralization. *Am J Pathol* 1999; 154: 671–5.
- [9] Lack EE, Young RH, Scully RE. Pathology of ovarian neoplasms in childhood and adolescence. *Pathol Annu* 1992; 27: 281–356.
- [10] Brammer IIIHM, Buck JL, Hayes WS, Sheth S, Tavassoli FA. From the archives of the AFIP. Malignant germ cell tumors of the ovary: radiologic-pathologic correlation. *Radiographics* 1990; 10: 715–24.
- [11] Levitin A, Hailer KD, Cohen HL, Zinn DL, O'Connor MC. Endodermal sinus tumor of the ovary imaging evaluation. *AJR Am J Roentgenol* 1996; 167: 791–3.
- [12] Thoshihide Y, Kaori T, Takashi K, *et al.* Yolk sac tumor of the ovary: radiologic–pathologic correlation in four cases. *J Comput Assist Tomogr* 2000; 4: 605–9.
- [13] Nawa A, Obata N, Kikkawa F, *et al.* Prognostic factors of patients with yolk sac tumors of the ovary. *Am J Obstet Gynecol* 2001; 184: 1182–8. doi:10.1067/mob.2001.113323. PMID:11349186.
- [14] Tewari K, Cappuccino F, Disaia P, Berman ML, Manetta A, Kohler MF. Malignant germ cell tumors of the ovary. *Obstet Gynecol* 2000; 95: 128–33. doi:10.1016/S0029-7844(99)00470-6. PMID:10636515.