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CASE REPORT

Pseudo-Meigs' syndrome with elevated serum CA 125 secondary to struma ovarii: A case report.

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ABSTRACT

We report a rare case of pseudo-Meigs' syndrome caused by ovarian stroma ovarii. A 63-year-old woman presented with ascites, hydrothorax, and elevated serum levels of CA 125. These findings were interpreted as suggestive of a possible ovarian malignancy. Thoracocentesis was performed pre-operatively due to her dyspnea. Pleural fluid was negative for malignant cell. Intraoperative frozen-section examination of the left ovary showed benign pure struma ovarii. A total hysterectomy with bilateral salpingo-oophorectomy and appendectomy were performed. Complete resolution of symptoms was achieved and the serum levels of CA 125 returned to normal after the operation.

Key Words: Pseudo-Meigs' syndrome, CA 125, struma ovarii

Meigs' syndrome is defined as benign and solid ovarian tumors with the gross appearance of a fibroma (fibroma, thecoma, and granulosa cell tumor), accompanied by ascites and hydrothorax, on condition that excision of this benign tumor cured the patient.[1] Pseudo-Meigs' syndrome refers to the same clinical features associated with other benign cysts of the ovary, ovarian metastasis from gastrointestinal cancer, leiomyomas of the uterus, and teratoma.[2-6] Stroma ovarii rarely presents as pseudo-Meigs' syndrome.[5, 7-12] We report a case of struma ovarii associated with ascites, hydrothorax, and elevated serum levels of CA 125 which may lead to suggestive of a possible ovarian malignancy.

Case report

A 63-year-old P6-0-1-5 woman was admitted to Prince of Songkla hospital after several months of abdominal distension and progressive dyspnea. She denied a history of a pelvic mass or any gastrointestinal symptoms. Her past history and family history were unremarkable.

Physical examination revealed dullness on percussion with decreased breath sounds in the upper two-thirds of the right lung field. Chest X-ray confirmed massive right pleural effusion (Fig. 1). Abdominal examination revealed marked distension, and shifting dullness consistent with significant accumulation of ascites. No abdominal or pelvic mass was palpable. However, a transabdominal ultrasound of the abdomen and the pelvis revealed a 6.9x5.5x5.0

cm mixed solid and cystic left adnexal mass with a large amount of ascites. The serum CA 125 level was 914 U/ml (normal value: below 35 U/ml). Other hematological and biochemical tests were normal.

On admission, she underwent thoracentesis to relieve dyspnea. Pleural fluids recurred rapidly and required retained intercostal drainage. The pleural fluid was negative for malignant cells. Five days later the patient underwent exploratory laparotomy. The preoperative diagnosis was an advanced stage of ovarian malignancy.

Operative findings revealed 10 liters of serous fluid, and a complex left ovarian mass measuring 6.0x4.0x4.0 cm (Fig. 2). This ovarian tumor was removed and sent for frozen section assessment. The uterus and right ovary were normal with no evidence of intraperitoneal or retroperitoneal disease. The frozen section reported a benign struma ovarii. A total hysterectomy with bilateral salpingo-oophorectomy and appendectomy was performed. The peritoneal fluids were also sent for cytologic examination.

Pathology revealed a pure stroma ovarii of the left ovary (Fig. 3). There was no evidence of malignancy in the specimens and peritoneal fluids were negative for malignant cells.

The post-operative course was uneventful. The patient recovered with no evidence of reaccumulation of pleural effusions or ascites (Fig. 4). Thyroid function tests performed postoperatively were within normal limits. One month after surgery the serum levels of CA 125 had fallen to normal.

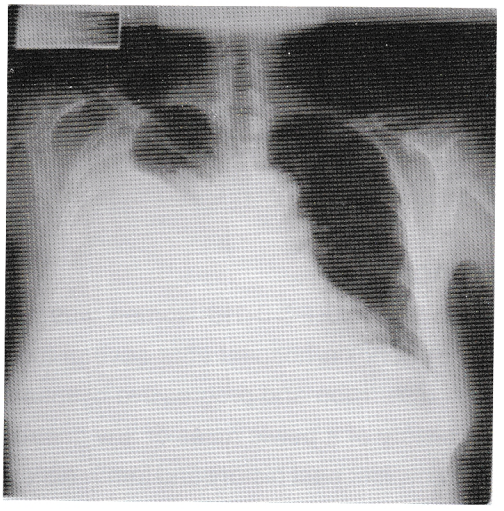

Fig. 1. Chest X-ray shows large right pleural effusion 2 days before surgery.

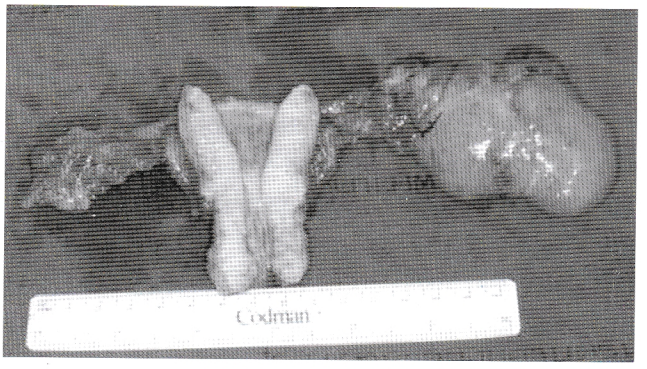

Fig. 2. Photograph of the uterus and bilateral adnexa. Left ovarian tumor is observed.

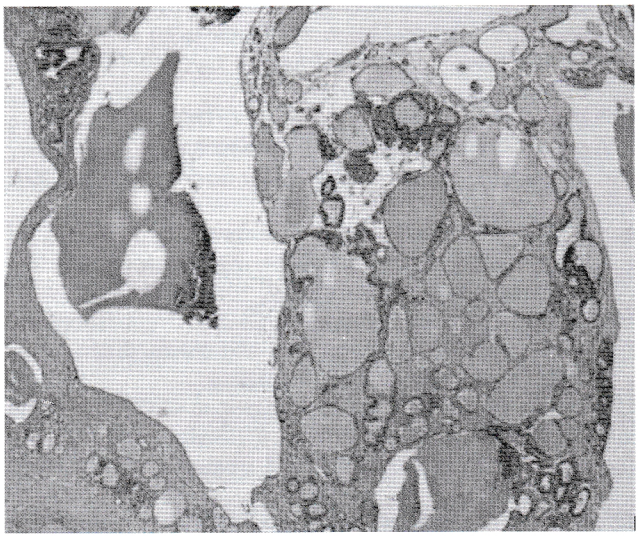

Fig. 3. Microscopic appearance of the left ovary showing various sizes of thyroid follicles.(H&E, low power)

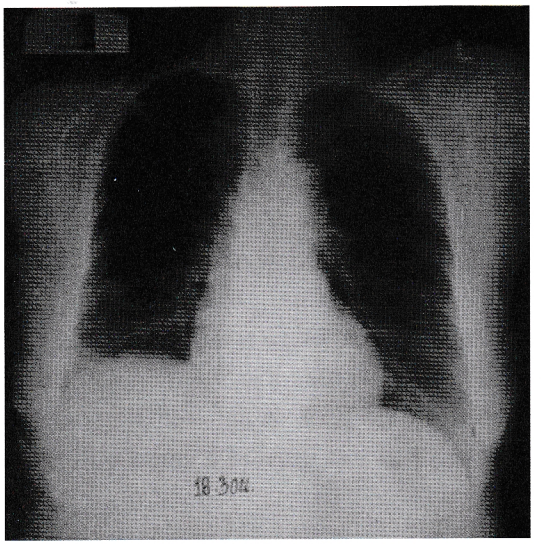

Fig. 4. Chest X-ray 7 days after surgery.

Discussion

Stroma ovarii was first described by Van Kalden in 1895 and Gottschalk in 1899.[13] It is a rare form of ovarian neoplasm, accounting for approximately 1% of all ovarian neoplasms.[7,14] Thyroid tissue is present in approximately 10% of mature teratomas and the tumor is described as a struma ovarii when it constitutes greater than 50% of the tumor.[7] Despite the predominant of thyroid tissue, only 5-8% of patients with this tumor have clinical features of hyperthyroidism.[7,13]

Patients with stroma ovarii usually present with abdominal or pelvic symptoms of a mass. Some cases have struma ovarii associated with ascites and hydrothorax.[5,7-12] Recently some authors have reported cases of this tumor associated with ascites, hydrothorax, and elevated serum CA 125 levels[7-12], as in this case.

Several mechanisms have been proposed to explain the origin of ascites and hydrothorax in Meigs' syndrome and pseudo-Meigs' syndrome. These include transudation of interstitial fluid, cyst formation within the tumor secondary to injury or necrosis,[15] pressure on the lymphatics in the tumor,[4] physical irritation of the peritoneum,[16] and twisting of the pedicle of the tumor and its torsion resulting in fluid production.[17] The formation of pleural fluid arises through direct passage of ascitic fluid from developmental defects in the diaphragm, and the greater proportion of right pleural effusions in Meigs' syndrome is due to the greater frequency of such defects on the right diaphragm.[6]

The presence of an ovarian mass with suspicious clinical signs and elevated levels of CA 125 may be suggestive of ovarian malignancy. CA 125 is a biochemical marker which has been shown to be of use in surveillance of patients with epithelial ovarian cancers. Overall, approximately 85% of patients with epithelial ovarian cancer have CA 125 levels above 35 U/ml.[18] The low positive predictive value of CA 125 for ovarian cancer in patients presenting with pelvic mass is due to the marker being elevated in other benign conditions including ovarian tumors.[15] The cause of the elevation of CA 125 levels may well be secondary to the presence of ascites.[9,11]

This case is an unusual case of benign struma ovarii associated with a large amount of ascites, hydrothorax, elevated serum CA 125 level and pelvic

mass in a postmenopausal woman, all suggesting an advanced ovarian cancer. Surgical excision of the ovarian tumor effected immediate and dramatic resolution of the ascites and hydrothorax.

In conclusion, although ovarian struma ovarii associated with ascites, hydrothorax, and elevated serum CA 125 level is rare, it should be included in the differential diagnosis of clinical features suspicious of ovarian malignancy. It is essential that a microscopic examination be made for accurate diagnosis, which will ensure correct management that will provide a good outcome with fewer complications.

References

1. Meigs JV. Fibroma of the ovary with ascites and hydrothorax; Meigs' syndrome. Am J Obstet Gynecol 1954; 67: 962-87.
2. Ohsawa T, Ishida H, Nakada H, Inokuma S, Hashimoto D, Kuroda H, et al. Pseudo-meigs' syndrome caused by ovarian metastasis from colon cancer: report of a case. Surg Today 2003; 33: 387-91.
3. Nagakura S, Shirai Y, Hatakeyama K. Pseudo-Meigs' syndrome caused by secondary ovarian tumors from gastrointestinal cancer. A case report and review of the literature. Dig Surg 2000; 17: 418-9.
4. Jimerson SD. Pseudo-Meigs' syndrome, an unusual case with analysis of the effusions. Obstet Gynecol 1973; 42: 535-7.
5. Morrell ND, Frost D, Ziel HK. Pseudo Meigs' syndrome. A case report. J Reprod Med 1980; 25: 88-90.
6. Bridgewater JA, Rustin GJ. Pseudo-Meigs' syndrome secondary to an ovarian germ cell tumor. Gynecol Oncol 1997; 66: 539-41.
7. Bethune M, Quinn M, Rome R. Struma ovarii presenting as acute pseudo-Meigs syndrome with an elevated CA125 level. Aust N Z J Obstet Gynaecol 1996; 36: 372-3.
8. Mancuso A, Triolo O, Leonardi I, De Vivo A. Struma ovarii: a rare benign pathology which may erroneously suggest malignancy. Acta Obstet Gynecol Scand 2001; 80: 1075-6.
9. Loizzi V, Cappuccini F, Berman ML. An unusual presentation of struma ovarii mimicking a malignant process. Obstet Gynecol 2002; 100: 1111-2.
10. Huh JJ, Montz FJ, Bristow RE. Struma ovarii associated with pseudo-Meigs' syndrome and elevated serum CA 125.Gynecol Oncol 2002; 86: 231-4.
11. Leung YC, Hammond IG. Limitations of CA 125 in the preoperative evaluation of a pelvic mass: struma ovarii and ascites. Aust N Z J Obstet Gynaecol 1993; 33: 216-7.
12. Bokhari A, Rosenfeld GS, Cracchiolo B, Heller DS. Cystic struma ovarii presenting with ascites and elevated CA-125 level. A case report. J Reprod Med 2003; 48: 52-6.
13. Dardik RB, Dardik M, Westra W,Montz FJ. Malignant struma ovarii: two case reports and review of the literature. Gynecol Oncol 1999; 73: 447-51.
14. Ayhan A, Yanik F, Tuncer R, Tuncer ZS, Ruacan S. Struma ovarii. Int J Gynecol Obstet 1993; 42: 143-6.
15. Timmerman D, Moerman P, Vergote I. Meigs' syndrome with elevated serum CA 125 levels: two case reports and review of the literature. Gynecol Oncol 1995; 59: 405-8.
16. Lin JY, Angel C, Sickel JZ. Meigs syndrome with elevated serum CA 125. Obstet Gynecol 1992; 80: 563-6.
17. Kempers RD, Dockerty MB, Hoffman DL, Bartholomew LG. Struma ovarii-ascites, hyperthyroid and asymptomatic syndrome. Ann Intern Med 1970; 72: 883-93.
18. Canney PA, Moore M, Wilkinson PM, James RD. Ovarian cancer antigen CA-125: a prospective clinical assessment of its role as a tumour marker. Br J Cancer 1984; 50: 765-9.