PSEUDO-MEIGS’ SYNDROME WITH DEGENERATIVE UTERINE LEIOMYOMA IN PREGNANCY

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SUMMARY

Objective: Uterine leiomyomas rarely cause pseudo-Meigs’ syndrome. We describe a case of pseudo-Meigs’ syndrome during pregnancy.

Case Report: A 35-year-old woman (gravida 0, para 0) experienced rapid abdominal distension over a period of 1 week in the first trimester. She had had an adnexal mass of 7 × 6 cm for 5 years but continued to have regular menstrual cycles. Serial imaging showed slow progressive enlargement of the adnexal mass but serum CA-125 levels remained normal. Ultrasonography showed a large solid mass that was 25 × 20 × 20 cm in size, with a uterine gestational sac and massive ascites. Both ovaries appeared normal and a small amount of pleural effusion was detected. CA-125 levels increased to 558.11 kU/L and exploratory laparotomy was performed. During surgery, the solid mass was found to be a pedunculated leiomyoma originating from the uterine fundus, with 3,200 mL of clear yellowish ascites. Myomectomy was performed and microscopic examination showed a leiomyoma with degenerative changes. The patient received regular prenatal examinations in outpatient clinics after discharge; a healthy female baby was delivered at term.

Conclusions: Pseudo-Meigs’ syndrome was diagnosed when hydrothorax and ascites occurred with a pelvic mass other than ovarian fibroma. Pregnancy may aggravate the syndrome. [Taiwanese J Obstet Gynecol 2004; 43(3):161–164]

Key Words: leiomyomas, pseudo-Meigs’ syndrome, pregnancy

Introduction

Uterine leiomyomas with ascites and hydrothorax were first reported in 1909 [1]. The significance of hydrothorax and ascites in patients with benign ovarian fibromas was further discussed in 1937 [2]. Meigs’ syndrome was categorized in 1954 as a benign solid ovarian tumor with the gross appearance of a fibroma (fibroma, thecoma, or granulosa cell tumor) accompanied by ascites and hydrothorax, which resolve after removal of the benign tumors [3]. Other benign cysts or teratomas of the ovary and leiomyomas of the uterus associated with ascites and hydrothorax are designated pseudo-Meigs’ syndrome [4]. Although leiomyomas are the most common benign tumors of the uterus, uterine leiomyomas are rarely accompanied by ascites and hydrothorax [5]. After searching the English language medical literature [6–9], we present the first case of pseudo-Meigs’ syndrome caused by a uterine leiomyoma with pregnancy in association with an elevated serum CA-125 level.

Case Report

A 35-year-old woman (gravida 0, para 0) experienced
rapid abdominal distension over the course of 1 week. She arrived at our outpatient clinic and reported being 11 days late for the start of her menstrual period. She had been having regular menstrual cycles and had never undergone surgery. An adnexal mass had been noted 5 years previously, for which she attended regular follow-up. The mass gradually increased from $7 \times 6$ cm to $13 \times 10$ cm between 1998 and 2002. However, the patient’s levels of CA-125, a serum tumor marker, had remained normal during this period. During the initial examination, ultrasonography showed a large solid mass ($25 \times 20 \times 20$ cm) with a uterine gestational sac comparable to 5 weeks’ gestation. In addition, massive ascites and some pleural effusion were noted (Figure 1). Both ovaries appeared normal. CA-125 level was 558.11 kU/L (normal, < 35 kU/L) and increased to 1,511.15 kU/L after 5 days. Chest roentgenogram was not obtained due to her early pregnancy.

Exploratory laparotomy was performed. A soft solid mass originating from the uterine fundus was discovered (Figure 2). The tumor was tan, elastic, and freely movable. The upper abdomen was free of disease and tumor excision was performed (Figure 3). Clear yellowish ascitic fluid (3,200 mL) was drained. Microscopic examination revealed that the mass was a benign mesenchymal tumor composed mainly of rounded epithelial cells with a minor spindling component (Figure 4). The stroma was well vascularized, comprising variable-sized, capillary-like to ectatic, hyalinized vessels. Leiomyma with degenerative change was diagnosed.

The patient was discharged after an uneventful recovery. She continued to receive regular prenatal examinations in outpatient clinics; a healthy female baby was delivered at term.

**Discussion**

Pseudo-Meigs’ syndrome is defined as the simultaneous occurrence of hydrothorax, ascites, and pelvic tumor other than an ovarian fibroma. Most of these cases involve ovarian pathologies and are rarely associated
with uterine leiomyomas [10]. For the few reported cases of pseudo-Meigs’ syndrome associated with leiomyoma [6], the average diameter of the leiomyoma was greater than 15 cm and the amount of ascitic fluid ranged from 1,600 mL to 19,000 mL (Table 1). The origin of the ascitic fluid in pseudo-Meigs’ syndrome remains uncertain. Some investigators believe that the fluid is transuded from the tumor surface. Others have proposed that ascites is produced either directly from the surrounding lymphatics or vessels due to tumor compression, hormonal stimulation, or tumor torsion [14]. The compression and subsequent congestion of lymphatic and blood vessels within the tumor or from the interstitial site of the tumor surface may also cause ascites [5]. In our case, the myometrial cysts contained a large amount of serous fluid, suggesting that fluid leakage from the cystic degenerated myoma, which might result from hydropic degeneration [15], was the source of the massive ascites. Intratumoral pressure might squeeze fluid in the cyst through intercellular spaces into the peritoneal cavity. In addition, mechanical tumor irritation and increased peritoneal pressure from the ascites can cause further peritoneal fluid production through peritoneal inflammation [6]. Hydrothorax may be due to fluid leakage from the abdomen and, after a sufficient volume and pressure is attained, through the diaphragm and into the intercellular gaps [16,17]. Hydrothorax could not be confirmed in our case because chest roentgenogram was not performed due to the patient’s pregnancy, but some pleural fluid was found while performing abdominal sonography.

Elevation of serum CA-125 together with a leiomyoma has been reported inconsistently, and the rise in CA-125 is usually small [18]. Peritoneal mesothelium has been reported as the source of elevated CA-125 in Meigs’ syndrome [19,20]. When present in pseudo-Meigs’ syndrome, CA-125 elevation is reportedly due to peritoneal inflammation, which has also been suggested as the source of ascites formation [6,18]. No differences in CA-125 levels are usually found between pseudo-Meigs’ syndrome and Meigs’ syndrome [21]. Table 1 shows the CA-125 levels reported in cases of pseudo-Meigs’ syndrome caused by a leiomyoma.

Only three cases of pseudo-Meigs’ syndrome associated with pregnancy have been reported; all were associated with ovarian tumors (Table 2) [7–9]. However, pseudo-Meigs’ syndrome caused by a degenerative myoma during pregnancy has not been reported. Our patient had undergone regular follow-up examinations for 5 years with no recurrence of ascites or elevation of CA-125 levels. The rapid elevation in CA-125 level and massive ascites formation immediately after conception suggests the development of pseudo-Meigs’ syndrome after degenerative changes in the leiomyoma associated with early pregnancy. The stimulatory effect of pregnancy on the growth of uterine myomas has been noted clinically and is assumed to be due to the presence of estrogen and progesterone receptors in the uterine and myomatous tissues [22]. Apart from elevated estrogen and progesterone secretion during pregnancy, which may cause myomatous degenerative changes, this process probably involves a more complex mechanism. Using serial ultrasound monitoring, Lev-Toaff and co-workers observed that myoma sizes either remained unchanged or increased during pregnancy, while larger myomas tended to

| Table 1. Characteristics of cases with pseudo-Meigs’ syndrome caused by uterine leiomyoma |
|-------------------------------------|--------|-----------------|-----------------|-----------------|
| Authors                            | Year   | Tumor size (cm) | Amount of ascites (mL) | CA-125 (kU/L) |
| Ollendorf et al [10]              | 1997   | 27 × 18 × 13    | 19,000                       | 301            |
| Dunn et al [13]                   | 1998   | 15 × 30 × 18      | 1,600                        | 254            |
| Amant et al [6]                   | 2001   | 30 × 30 × 15      | 12,000                       | 785            |
| Hsu et al [this paper]            | 2004   | 25 × 20 × 20      | 3,200                        | 1,511          |

NA = not available.

| Table 2. Characteristics of cases with pseudo-Meigs’ syndrome and pregnancy |
|-------------------------------------|-----------------|-----------------|-----------------|-----------------|
| Authors                            | Gestational age at diagnosis | Tumor type                    | Pregnancy outcome |
| Gorsse [7]                         | 2.5 months’ gestation | Pseudomucinous cystadenoma | Term delivery   |
| Printer [8]                        | Near term          | Pseudomucinous cystadenoma | Term delivery*  |
| Jimerson [9]                       | Postpartum         | Pseudomucinous cystadenocarcinoma | Term delivery*  |
| Hsu et al [this paper]             | 6 weeks            | Leiomyma                   | Term delivery   |

*Laparotomy after delivery.
become smaller during the second trimester [23]. Although a gradual increase in the levels of plasma progesterone, estradiol, and estriol occur in normal pregnancy, it is likely that estrogen receptors in myomatous tissues are down-regulated later in pregnancy due to the large amount of secreted estrogen. Therefore, myoma size usually remains unchanged or decreases in the third trimester [22,23].

Leiomyoma is the most common benign tumor of the uterus and may be submucosal, intramural, or subserosal in location. Subserosal leiomyomas are either beneath the uterine surface or exophytic into a pedunculated form. A pedunculated subserosal leiomyoma has been called a “migrating” or “wandering” leiomyoma when the stalk is extremely long [24]. Occasionally, a leiomyoma is found separated from the uterus, and has acquired its vascular connections from the omentum, pelvic wall, or other intra-abdominal sites such as the cecal wall. In this case, it is termed a parasitic leiomyoma [24,25]. Pedunculated or parasitic leiomyomas may present with a wide spectrum of symptoms and, therefore, pose a diagnostic dilemma [10,26,27]. The pedunculated myoma in our case was misdiagnosed as an adenxal tumor. The association of pedunculated leiomyomas in pregnancy with pseudo-Meigs’ syndrome is a rare but clinically favorable disease after excision of the leiomyoma. Differential diagnosis of pedunculated myoma and pseudo-Meigs’ syndrome must be kept in mind when a solid pelvic mass associated with massive ascites, pleural effusion, and elevated levels of serum CA-125 is encountered.

References