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A case of retroperitoneal liposarcoma after delivery with expression of estrogen receptor: Report of a case

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ABSTRACT

INTRODUCTION: Liposarcoma is one of the most common soft tissue sarcomas; however, early diagnosis is rare as the tumor remains difficult and unpalpable for a prolonged period of time.
PRESENTATION OF CASE: Here we report the first case of retroperitoneal liposarcoma associated with pregnancy and expression of estrogen receptor. A 34-year-old woman experienced persistent abdominal distension after her first delivery. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed a large (40 cm × 35 cm), solid, palpable abdominal mass with fat attenuation displacing the ascending colon and the right kidney to the left. Laparotomy and an en-bloc resection of the tumor were performed; further, right nephrectomy and adrenalectomy were required. Histopathology showed a well-differentiated liposarcoma; approximately 10–20% of the tumor cells were ER-positive.
DISCUSSION: Retroperitoneal liposarcoma associated with pregnancy is an extremely rare occurrence. Surgical resection is unquestionably the first choice of treatment, but complete resection is sometimes impossible due to the volume and depth of invasion of the tumor. In such cases, additional therapy for liposarcoma is important to improve prognosis. Thus, this report highlights the need for further research into hormone therapy.
CONCLUSION: Retroperitoneal liposarcoma has a high local recurrence rate due to the difficulty in complete surgical resection; therefore, additional hormone therapy is important for improving the prognosis.

1. Background

Liposarcoma is one of the most common soft tissue sarcomas, yet early diagnosis is rare because the tumor remains difficult and unpalpable for a prolonged period of time. The peak incidence of liposarcoma occurs from 40 to 60 years of age with the preponderance of cases reported in men [1]. It is very rare in females of reproductive age, and the prognosis is better in pre-than in post-menopausal women [2]. Although some reports mention an association between steroid hormones and malignant soft tissue tumors including liposarcomas, a case of liposarcoma associated with pregnancy wherein hormone receptors have been investigated has not been previously reported. Here, we report the first case of retroperitoneal liposarcoma associated with pregnancy and ER expression and review the literature.

2. Case report

A 34-year-old woman experienced persistent abdominal distension for the duration of her pregnancy with no improvement even after delivery. She had a normal delivery eight months earlier, and her child displayed normal growth. Although no abnormalities were detected during the several routine transvaginal ultrasonographic examinations performed during pregnancy, at birth, and six months after delivery, contrast enhanced computed tomography (CT) and magnetic resonance imaging (MRI) (fat suppression, T1WI) revealed a 40 cm × 35 cm solid mass with fat attenuation displacing the ascending colon and the right kidney to the left (Fig. 1A and B). Furthermore, this was accompanied by breathlessness due to restriction of diaphragmatic movement. Laboratory findings including tumor marker CA-125, alpha-fetoprotein, and carcino-embryogenic antigen levels were within the normal range. Laparotomy was performed under general anesthesia, and a mass was found to be wrapped in the retroperitoneum and pseudomembrane. The uterus, uterine tube and ovaries were normal. We performed an en-bloc resection of the tumor, along with right nephrectomy and adrenalectomy.

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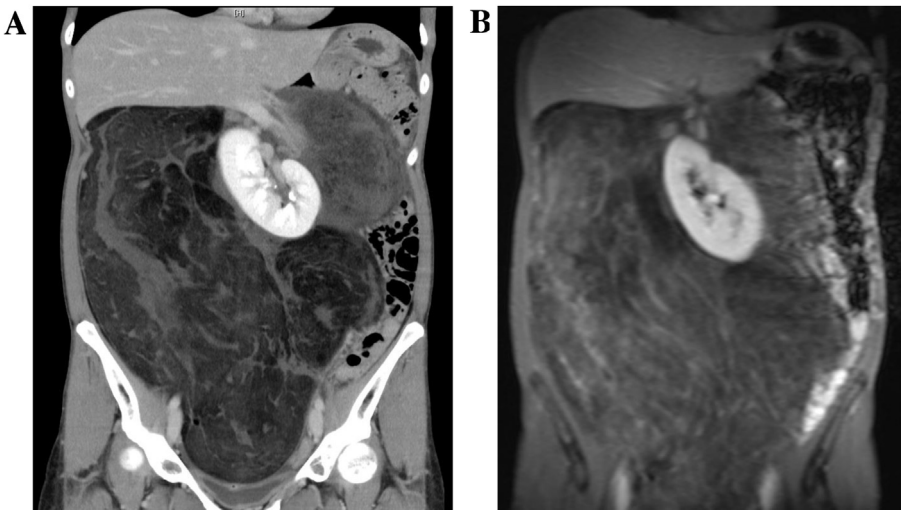

Fig. 1. (A) contrast enhanced CT, (B) MRI (fat suppression, T1WI) A 40 × 35 cm solid mass with fat attenuation displacing ascending colon and right kidney to left.

The retroperitoneal mass weighed 7200 g and measured 43 cm × 40 cm × 13 cm (Fig. 2). Histopathological examination of the surgical specimen showed a well-differentiated liposarcoma (Fig. 3). Immunohistochemistry revealed approximately 10–20% of the tumor cells to be estrogen receptor (ER)-positive, with negative results for the progesterone receptor (PR) (Fig. 4). The patient recovered uneventfully and was discharged from the hospital on the 15th postoperative day. She and her husband were planning to try for another baby, so postoperative adjuvant therapy including hormone therapy was not undertaken. No recurrence was detected at the 3-year follow-up.

3. Discussion

Retroperitoneal sarcomas represent approximately 10–15% of all soft tissue sarcomas, which themselves are rare, accounting for only 1% of all malignancy [3]. The peak incidence of liposarcoma occurs from 40 to 60 years of age. The tumor is typically aggressive, with reported 5-year survival rates ranging from only 23% to 46% [4]. Lewis et al. [5] reported high local recurrence rates for retroperitoneal sarcoma, ranging from 40% to 80%. As the retroperitoneal space is easily distended by the enlarging tumor, the neoplasia remains inactive and unpalpable for prolonged period of time. A retroperitoneal mass may produce a wide range of signs and symptoms due to the compression and infiltration of the surrounding organs: localized pain associated with neurologic symptoms due to neuroplexus involvement, vomiting due to gastric compression, constipation, hydronephrosis, uremia due to renal disorder, and ureteric compression. Izumi et al. [6] reported that the most common symptom of retroperitoneal sarcomas is abdominal pain (25.8%), followed by abdominal distension and palpable mass, while 22.1% of the cases develop without symptoms. In the present case, the first symptoms were continuous abdominal distension and respiratory discomfort lasting throughout the pregnancy without improvement, suggesting that the mass existed and grew during the pregnancy. Ultrasonographic examinations performed as routine check-ups during the perinatal period did not detect the retroperitoneal tumor in this case. Hence, it is recommended that a transabdominal ultrasonographic examination (or MRI if necessary) be performed in cases where a pregnant woman complains of abdominal distention or respiratory discomfort.

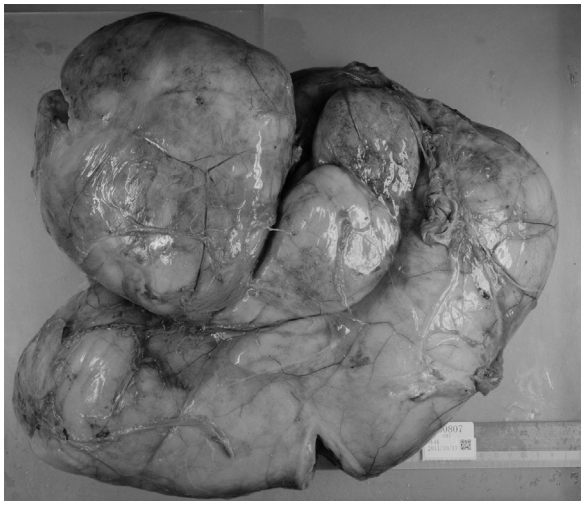

Fig. 2. Gross appearance of the tumor. An elastic soft mass covered pseudocapsule weighed 7200 g and measured 43 cm × 40 cm × 13 cm with lobulations.

We reviewed the literature on liposarcoma associated with pregnancy and found 16 cases including ours. Their clinical courses are summarized in Table 1 [1,3,7,8,10–18]. The mean age was 31 years (range, 15–44 years). Four cases of liposarcoma were discovered during the first trimester, 10 during the second or third trimester, and 2 within the 1 year postnatal period. Only 2 patients diagnosed in the first trimester were surgically treated during pregnancy; in all other cases, surgery was performed after delivery.

Liposarcoma is classified into five histological subtypes: well-differentiated, round-cell, myxoid, pleomorphic, and de-differentiated (WHO classification, 2002). There were 8 cases of myxoid type and 4 cases of well-differentiated type, both of which were classified as low-grade. Twelve cases were large tumors exceeding 15 cm in diameter. According to the reports, surgery was the only treatment given during pregnancy; however, radiotherapy to the lower extremity after delivery [7] and chemotherapy in combination of mesna, doxorubisin and ifosfamid after surgical excision [8] have been reported. In terms of prognosis, out of the 4 patients diagnosed in the first trimester, 2 survived following surgery under general anesthesia at 13th week of pregnancy. However, all patients for whom surgery was performed after delivery died. Therefore, surgical resection should be considered as soon as possible, even if the initial diagnosis is made during pregnancy.

The effect of pregnancy on tumors is a major concern. In our patient, a relationship existed between tumor growth and pregnancy, and immunostaining was ER-positive. Cantin and