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An Unusual Case of Dysgerminoma and Review of the Literature

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ABSTRACT

Malignant germ cell tumour is a rare form of cancer which affects 0.07% of woman globally and they usually occur in the ovaries. Extragonadal sites for germ cell tumours can occur in or near the midline, with the anterior mediastinum being the most common site.

Dysgerminoma is a form of malignant germ cell tumour and extragonadal sites in the retroperineum and anterior mediastinum, albeit rare, have been reported. There has not been any report of dysgerminoma arising from any intraperitoneal structures except for the ovaries to our best knowledge.

We present an unusual case of dysgerminoma presenting as torsion of a mass around a fallopian tube in the presence of two normal ovaries. We discuss dysgerminomas and unusual locations of ovaries and their associated pathology in this report.

Keywords: Dysgerminoma, ectopic ovary, torsion, accessory ovary, germ cell tumour

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INTRODUCTION

Malignant germ cell tumour is a rare form of cancer which affects 0.07% of woman globally[1] and they usually occur in the ovaries. Extragonadal sites for germ cell tumours have been reported and the most common site is the anterior mediastinum[2]. Dysgerminoma is a form of malignant germ cell tumour and extragonadal sites in the retroperineum and anterior mediastinum, albeit rare, have been reported[3]. A literature search using the keywords dysgerminoma, ectopic ovary, accessory ovary, torsion and germ cell tumour was carried out and there has not been any report of dysgerminoma or malignant germ cell tumour arising from any intraperitoneum structure except for ovaries to our best knowledge.

We present an unusual case of dysgerminoma arising in the pelvis in the presence of normal ovaries and the relevant literature review.

CASE PRESENTATION

A 17-year old woman presented to the emergency room with a history of progressive worsening lower abdominal pain of two months and a sudden exacerbation of pain for two hours. On examination, there was left iliac fossa tenderness with signs of acute abdomen. CT abdomen and pelvis showed a large lobulated mildly enhancing 13 x 12 cm soft tissue density solid mass lesion arising from the pelvis with moderate amount of free peritoneal fluid suggestive of haemoperitoneum. Ultrasound of the pelvis showed a large left adnexal mass demonstrating attachment to a vascular pedicle, suggesting torsion of a pedunculated appendage with haemoperitoneum present. CA 125 was mildly elevated at 55.6U/ml. The other ovarian tumour (AFP, CEA and βhCG) markers were normal.

She underwent laparoscopic left salpingectomy and removal of the adnexal mass on the same day. Intraoperatively, there was a left para-ovarian mass measuring 20x20cm (Figure 1), which had torted around the left fallopian tube thrice. The left tube was gangrenous. Both ovaries and the right tube were normal. There was one litre of haemoperitoneum. The mass was too large to fit into the lap sac bag for retrieval laparoscopically thus a mini-laparotomy was performed over the right side port to remove the mass from the abdomen. The mass appeared distinctively separate from the left ovary (Figure 2).

Her post-operative recovery was uneventful and she was discharged well on the second post-operative day. Histological examination showed a dysgerminoma comprising sheets of pleomorphic round cells immunoreactive for PLAP (Figure 3), CD117 and D2-40, interposed by fibrous bands containing mature lymphocytes. Focal haemorrhagic infarction indicative of torsion was also present (Figure 4).

Although a full surgical staging was not done at the time of the operation, post-operative CT scan did not have any evidence of intra-thoracic or intra-abdominal malignancy. She was staged as 1a dysgerminoma according to FIGO classification and is on close observation with regular serum lactate dehydrogenase measurement and ultrasound of the pelvis.

DISCUSSION

The para-ovarian mass tightly torted around the ipsilateral fallopian tube causing it to be gangrenous and the histological examination confirmed that the mass was loosely adhered to the fallopian tube. As there were two normal ovaries at the end of the surgery and there was no invasion of surface of the tube, the mass arose from something that was clearly separated from the ipsilateral ovary and fallopian tube. CT scan performed after surgery also confirmed the presence of two normal ovaries. The risk of having a malignancy presenting as torsion in a young patient is very low i.e. 1- 2%[4] thus the diagnosis of dygerminoma was a surprise. Pre-operative serum LDH was not done because we did not anticipate this diagnosis. The other ovarian tumour markers were normal.

Women with malignant germ cell tumour have a favourable prognosis as they usually present early and respond well to treatment[1]. Fertility-sparing surgery with a unilateral salpingo-oopherectomy, peritoneal washings and thorough inspection of the abdominal cavity is now considered standard treatment in early stage germ cell tumour[5]. Surveillance with regular ultrasound can be done for patients with stage 1a dysgerminoma. Although relapse in patients with stage 1 dysgerminoma is around 15-25%, they are very sensitive to platinum-based chemotherapy and salvage rate is high[5]. Chemotherapy using bleomycin, etoposide and cisplatin (BEP) is considered gold standard as adjuvant treatment for women with higher stage disease[6]. Weinberg et al reported a 100% resumption of normal menstruation in all the patients following fertility-sparing surgery and chemotherapy and that fertility does not seem to be affected by the treatment[6]. Dysgerminoma is exquisitely sensitive to radiotherapy but pelvic radiation is associated with gonadal dysfunction and loss of fertility[5]. This patient was discussed at the tumour board in our institution and the decision was not to proceed with an interval salpingo-oopherectomy because the mass did not arise from the normally-placed ovary. It was in fact from a separate structure that was not part of the ovary or fallopian tube.

Extragonadal germ cell tumour usually occurs in the midline e.g. anterior mediastinum, sacrococcygeal region and the pineal region[7]. A literature review was carried out but we could not find any reports of dysgerminoma arising from the fallopian tubes or nearby structures in the peritoneal cavity, excluding the ovaries. We were unable to correlate the source of the mass with a usual anatomical structure in the pelvis but we found literature reporting benign and malignant tumours including serous

papillary carcinoma, mucinous cystadenocarcinoma and Brenner tumour arising from ectopic ovaries[8-9].

Ectopic ovary is a rare occurrence and its true incidence is unknown. They are often small and can be overlooked or mistaken as lymph nodes during surgeries[10]. Ectopic ovary behaves like a normally placed ovary and can respond to ovarian stimulation. In fact, stimulation with clomiphene citrate and looking for follicles has been used to confirm the diagnosis of ectopic ovaries[11].The estimated incidence is between 1in 29,000 and 1 in 700,000 gynaecological admissions but is often thought to be an underestimate[12].It has been reported in the pelvis, on the broad ligament, in the retroperitoneum near the para-aortic area, on the sigmoid colon, on the omentum and intrarenal presenting as a renal mass[8-9,13].

Terminology describing this occurrence has been confusing, creating much debate. It was first suggested by Wharton in 1959 to separate the entities of supernumerary ovary and accessory ovary by the location and blood supply of the ovarian tissue. A supernumerary ovary must be entirely separate from the normally located ovary whereas an accessory ovary is closely related to the normal ovary and is supplied by vessels continuous with those supplying the normal ovary[14]. Lachman et al. have suggested using the term ectopic in place of the traditional terms and categorizing them as post-surgical, post-inflammatory, or truly embryological as it has been documented that up to half of the women with additional ovaries have previous surgeries[15].

Congenital anomalies have been frequently reported in women with supernumerary and accessory ovaries. In 36% of reported cases, anomalies of the genitourinary tract, adrenal gland and liver have been documented[13]. We suggest screening all women with ectopic ovaries for associated anomalies especially in the genitourinary system as they occur most frequently there.

Both ectopic ovaries and dysgerminomas are rare events. We hope to emphasize that tumours typically occurring in ovaries can present in unusual place and manner and a high index of suspicion is needed.

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Figure 1: Paratubal mass upon entry into the abdomen

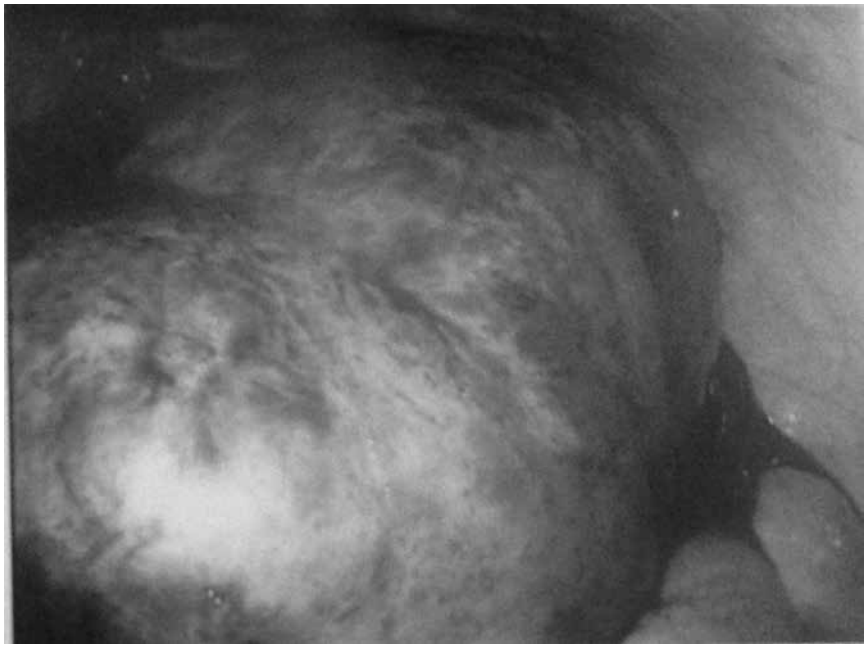

Figure 2: Both ovaries normal after removal of the mass

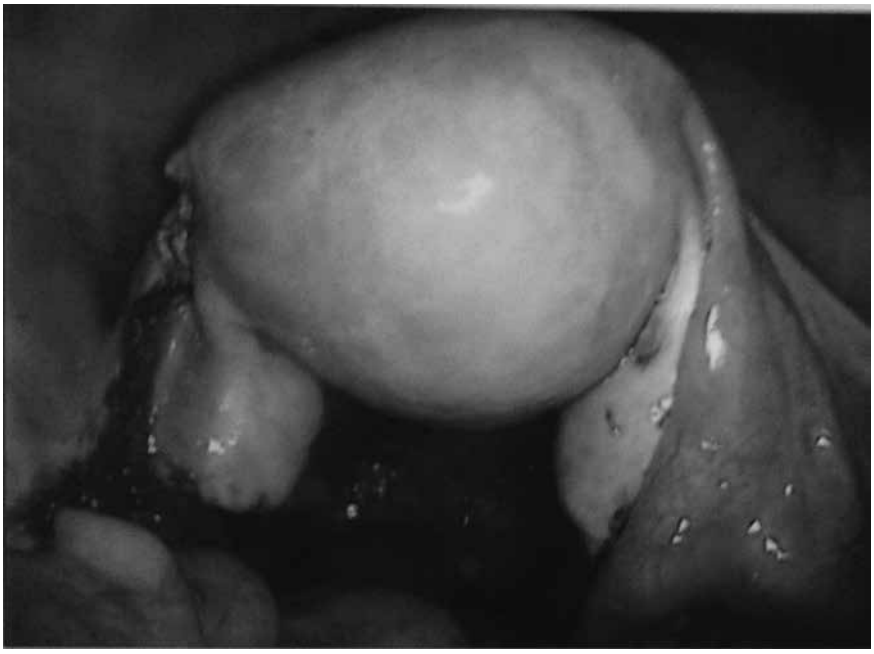

Figure 3: Tumour cells are immunoreactive for PLAP (x 400)

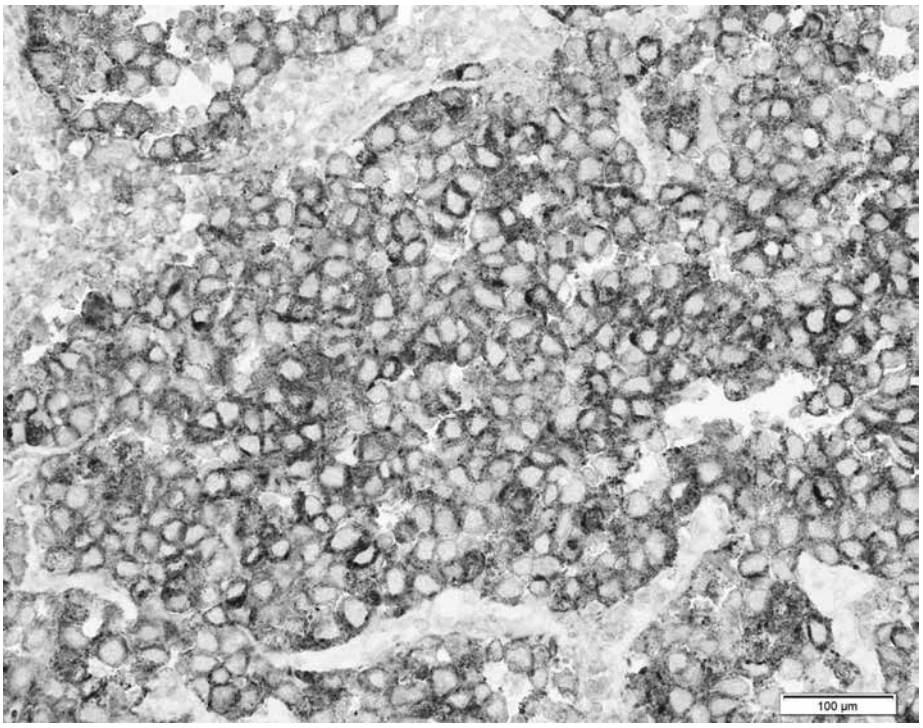

Figure 4: Dysgerminoma with sheets of round pleomorphic cells separated by fibrous bands with mature lymphocytes (H&E X 200).