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CASE REPORT

A Young Girl with Recurrent Fallopian Tube Carcinoma (FTC): An Interesting Case Report

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ABSTRACT

Primary fallopian tube carcinoma (FTC) is a rare disease which frequently occurs among post-menopausal women. It is often grouped under the epithelial ovarian cancer umbrella. The treatment of choice is surgery and chemotherapy. Our patient was a young teenage girl with recurrent FTC who responded well to surgery and chemotherapy. We discuss on the epidemiology, risk factors, principles of management and prognosis of FTC.

INTRODUCTION

Primary fallopian tube cancer (FTC) is one of the rare gynaecological malignancies. It accounts for 0.14% to 1.8% of female genital malignancies (Kalampokas et al., 2013). From literature, FTC was identified in the year 1847 but the first case report was published in 1888 by Orthmann. Its global incidence is 0.36-0.41 per 100,000 women per year (Marina et al., 2019). Around 40% to 60% of the tumours that are classified as high-grade serous carcinomas of the ovary or peritoneum might have originated from the fimbriae of the fallopian tube (Kindelberger et al., 2007). It is often associated with chronic tubal inflammation, infertility, tuberculous salpingitis and tubal endometriosis (Mladenovic, 2009). Similar to ovarian cancer, breast cancer 1 (BRCA 1) and tumour protein 53 (TP53) mutations are associated with fallopian tube malignancies (Senturk et al., 2010).

We report a case of a young girl who presented to us with gross ascites. An elective right salpingo-oophorectomy was performed. The histopathological examination (HPE) confirmed the diagnosis of right FTC. Despite on adjuvant chemotherapy and regular follow up, recurrence occurred after three years of remission. An extrafascial hysterectomy, left salpingo-oophorectomy, omentectomy, appendectomy and lymphadenectomy were done and she responded well to another cycle of carboplatin based chemotherapy.

CASE PRESENTATION

A healthy teenage virgin girl (weight = 60 kg, height = 1.7 metres), presented to us with a history of progressive abdominal distension for four weeks. It was associated with loss of appetite and weight for two weeks. She attained menarche at 11 years old and her menstrual cycle was normal and dysmenorrhea was not associated. She denied any history of trauma or fever. Her family history was unremarkable of any malignancies.

On examination, she was alert, conscious but cachexic. Her blood pressure (BP), heart rate (HR), respiratory rate (RR) and oxygen saturation (SaO2) were 112/75 mmHg, 92 beats per minute, 22 breaths per minute and 97%, respectively. Clinical examinations were unremarkable except for massive ascites.

INVESTIGATIONS

Her full blood count (FBC) revealed haemoglobin (Hb) of 10.2 g/dl (normal values: 11-13 g/dl), total white blood cells (TWBC) of 9.8 x 109/L (normal values: 7-12 x 109/L) and platelets of 273 x 109/L (normal values of 150-450 x 109/L). Other biochemical parameters, including the arterial blood gases (ABG), liver and renal function tests were within normal ranges. Tumour marker values such as carbohydrate antigen (CA) 19-9, carcinoembryonic antigen (CEA), alpha fetoprotein (AFP), lactate dehydrogenase (LDH), beta hCG and oestradiol were within normal values.

However, the cancer antigen 125 (CA-125) was grossly elevated at 457.1 U/ml (normal values: 0-35 U/ml). Ultrasound abdomen and pelvis revealed severe exudative ascites with mesenterial pseudocyst and omental thickening suggestive of tuberculous peritonitis with less probable differential diagnosis of peritoneal carcinomatosis. Peritoneal tapping was done and straw-coloured ascites fluid was sent for cytology, acid fast bacilli (AFB) and bacterial culture. The results were no malignant cell seen, no AFB seen, no bacterial growth respectively. Her computed tomography (CT) of thorax, abdomen and pelvis (TAP) showed extensive amount of ascites fluid and a cystic and vascularised lesion superior to the right side of uterus measuring 4.2 cm x 4.4 cm x 4.2 cm with thick and enhancing wall. There was a small cystic lesion at the left para adnexa region measuring 2.9 cm x 2.5 cm x 2.2 cm suggestive

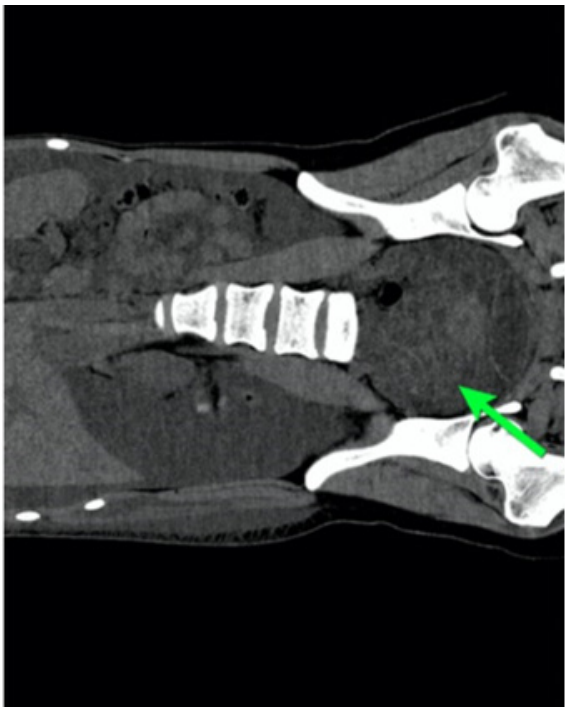

Figure 1: Coronal view of the pre-operative CT-TAP showing cystic and vascularised mass at the right adnexa.

of ovarian cystic mucinous adeno-carcinoma (Figure 1).

DIFFERENTIAL DIAGNOSIS

Based on the clinical findings and scan, a diagnosis of a huge right ovarian tumour was made. Epithelial ovarian tumour was our provisional diagnosis since CA-125 was elevated (in cases of serous epithelial ovarian tumour) and cystic appearance of the tumour in CT scan. As she was not sexually active with no history of fever, thus, tubo-ovarian mass or abscess (pelvic inflammatory disease) was excluded. She also did not have any history of dysmenorrhea, thus excluding the diagnosis of endometrioma. Dermoid cyst was another possible differential diagnosis, but cystic appearance of the tumour was not the common feature of dermoid cyst in CT scan

TREATMENT

Family conference was organised to discuss about prognosis of fallopian tube cancer and procedure before informed consent was taken. Our patient was posted for an elective laparotomy and excision of right ovarian tumour under general anaesthesia (GA). An epidural catheter was sited at the lumbar 3-4 (L3-4) vertebra level for perioperative analgesia. After that, she was intubated with a size 7 mm endotracheal tube and maintained with balanced anaesthesia which consisted of sevoflurane, morphine, rocuronium, and fractional of inspired oxygen (FiO2) of 40%. Ventilation was achieved with a peak airway pressure (PAP) of 29-32 cmH20 generating a tidal volume of 300-350 ml. Intraoperatively, uterus, the left fallopian tube and left ovary were noted to be healthy. However, there was a mass measuring 4.2 cm x 4 cm at the right fallopian tube with breached capsule, solid and irregular material seen. Tumour implant was seen at right anterior round ligament. Small intestine and large intestine were free from nodules. Urinary bladder was normal. A right salpingo-oophorectomy, omentectomy and peritoneal fluid sampling were performed and the samples were sent for histopathology examination (HPE).

She was extubated at the end of the surgery and sent to the intensive care unit (ICU) for monitoring and postoperative stabilization. She was discharged home four days later. Two weeks later, the HPE was reported as low-grade serous carcinoma of the right fallopian tube with invasive implant of low-grade serous carcinoma at the right round ligament. Theca luteal cyst was seen in right ovary. There was no malignancy on omentum. Our patient was staged with tumour, nodes, and metastasis (TNM) staging of TIIa N0 M0 which is equivalent to International Federation of Gynaecology and Obstetrics (FIGO) IIA. She was referred to the oncologist for chemotherapy.

Prior to chemotherapy, which commenced five weeks after the surgery, the CA-125 level decreased to 26.9 U/ml. She underwent a total of six cycles of paclitaxel-carboplatin chemotherapy regimen and the disease went into remission with the CA-125 level further dropped to 12.9 U/ml after six months. She was followed up closely in our gynaecological clinic with yearly CT TAP surveillance to monitor her disease progress.

However, three years later, she complained of intermittent lower abdominal pain for two months which was associated with loss of appetite and weight for one month. She also complained of abdominal distension with a left lower abdominal mass. On examination, the mass was palpable with a size of 8 cm x 6 cm at the left iliac fossa region. It was firm in consistency, mobile and there was no tenderness. Other clinical examinations were unremarkable. The CA-125 level was raised at 35.2 U/ml. Her repeated CT TAP showed an enlarged left adnexal complex cystic mass measuring 5.2 cm x 8.2 cm x 5.9 cm. However, there were no other localized or distant metastases seen (Figures 2 and 3).

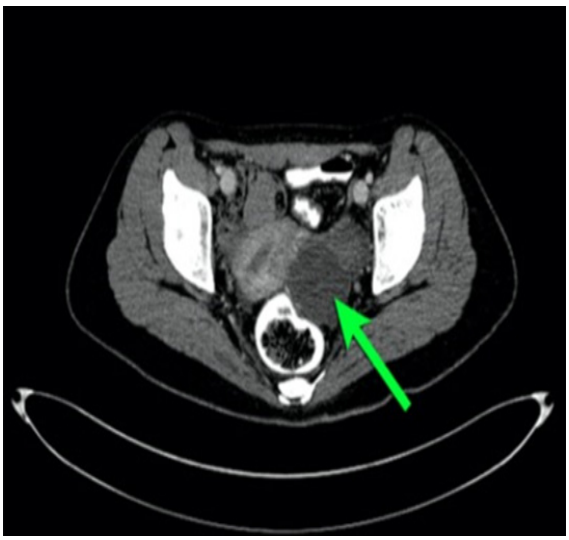

Figure 2: An axial view of CT-TAP which showed a huge left adnexal complex mass prior to the second surgery.

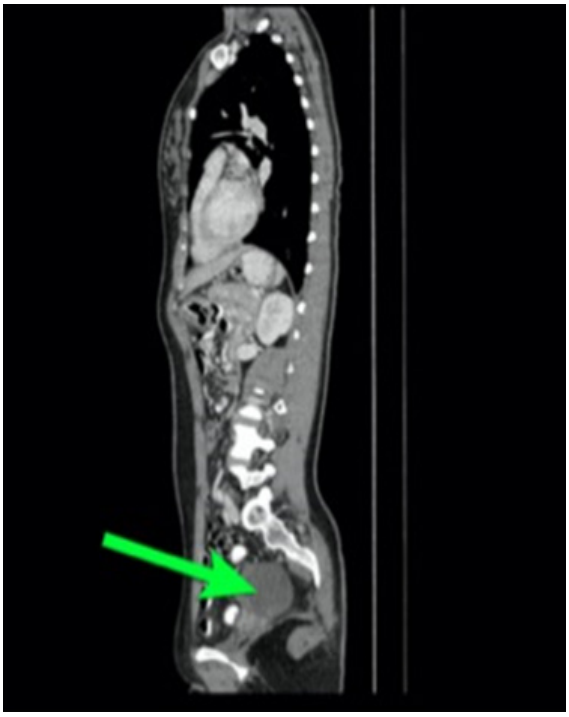

Figure 3: Sagittal view of CT-TAP prior to the second surgery.

Prior to elective operation, family conference was organised to discuss about fertility issue and advantages and disadvantages of hysterectomy. Family and patient were well aware of consequences and informed consent was taken. Oocyte harvesting was not done in view of the high cost. Genetic testing and molecular testing were not done pre-operatively. The option of hormonal suppression was not documented in case record She successfully underwent an elective laparotomy and left ovarian cystectomy three days later. The HPE of the left ovarian cyst revealed a serous borderline tumour with invasive implants on the uterus. A cycle of chemotherapy was urgently initiated after discussion with the oncologist. Recurrence of FTC was noted and the patient subsequently underwent an extra fascial hysterectomy, left salpingo-oophorectomy, omentectomy, appendectomy and lymphadenectomy.

OUTCOME AND FOLLOW-UP

Primary FTC is a rare gynaecological malignancy, accounting to 0.14-1.8 % of all gynaecological malignancy (Pectasides et al., 2006). It usually occurs in the postmenopausal elderly women with a mean age of 55 years old (age range of 17 to 88 years old) (Jeung et al., 2009). We believe that our unfortunate teenage girl was the youngest to be diagnosed to have stage II low grade serous FTC. Given the rarity of the disease, literatures on FTC and its recurrences are extremely limited.

The aetiology of this malignancy is unknown. High parity has been reported to be protective, and the use of oral contraceptives pills (OCP) decreases the risk of having FTC [Riska & Leminen, 2009). Theoretically, it is associated with tubal inflammation and endometriosis. Our patient was a nulliparous girl with no history of sexual relationship and nor consuming OCP.

Primary serous adenocarcinoma of the fallopian tube with papillary features is the commonest histological type. In comparison to ovarian carcinoma, FTC often presents at early stages, but with a worse prognosis. It is usually managed in the same manner as ovarian cancer (Kosary & Trimble, 2002).

In a retrospective study of 151 patients, it was shown that patients with FTC often present

with abnormal vaginal bleeding (47.5%), lower abdominal pain (39%), abnormal watery vaginal discharge (20%) and a palpable pelvic/abdominal mass (61%) (Baekelandt et al., 2000). Hundal et al. (2021) published a case of FTC in a pre-menopausal woman presenting with abnormal vaginal bleeding. Our patient initially presented with gross ascites, loss of appetite and weight. She did not present with abnormal vaginal bleeding and the other three symptoms. Therefore, it did not fit into the common clinical features of fallopian tube malignancy.

FTC can often be mistaken as tuberculous abdomen or salpingitis as tuberculosis is very common in Southeast Asian countries. Initial extensive radio-imaging results could not clearly reveal the diagnosis of primary FTC. The finding can mimic ovarian tumour, tubo-ovarian abscess in sonographic investigations (Haratz & Russell, 2004). Ovarian tumour diagnosis was even highlighted to be considered in the imaging report in our patient case. However, intraoperatively FTC diagnosis was confirmed. Therefore, operative findings predict the histological diagnosis, staging and prognosis (Berek et al., 2020).

Making the preoperative diagnosis could be assisted by measurement of the serum CA-125 levels, which is elevated in 80% of patients with FTC (Rezvani & Shaaban, 2011). It can also be raised in benign pelvic organ tumours such as endometriosis and pelvic inflammatory disease. Hefler et al. (2000) stated that median CA-125 level in patients with primary FTC preoperatively is 183 U/ml. Given that she was a young girl, FTC was not a likely diagnosis preoperatively despite a grossly elevated CA-125 (457.1 U/ml).

FTC spreads by local invasion, transluminal migration, lymphatics and haematological. It has a higher rate of retroperitoneal and distant metastases than that of epithelial ovarian cancer (Ajithkumar et al., 2005). The stage of disease at the time of diagnosis is the most important factor affecting the prognosis. Most of primary FTC cases are detected in early stage, in Taiwan >50% of patients are diagnosed to have stage I and II disease (Horng et al., 2014). Wethington et al. (2008) reported that the 5-year survival rate for Stage I tumour was 81%, and cancer-specific survival was 65% (95% CI 57–75) and 54% (95% CI 48–60) for Stages II and III, respectively. Survival rate for Stage I and II diseases has ranged from 37% to 95%, and for Stages III and IV tumour, from 0% to 69% (Wethington et al., 2008). The initial staging of our patient at the time of diagnosis was stage II, thus having a good survival rate.

The other clinicopathologic prognostic factors include residual disease after cytoreduction, the presence of ascites and the histological grading. Surgery is the treatment of choice, and the principles are the same as those used for ovarian cancer. Total hysterectomy, bilateral salpingo-oophorectomy, omentectomy, selective pelvic and para-aortic lymphadenectomy for any stage for FTC is commonly performed. Lymph node metastasis is common in patients with FTC, therefore lymphadenectomy is highly recommended (Koo et al., 2011). However, due to the age of our patient and fertility sparing factor, a laparotomy right salpingo-oophorectomy and omentectomy were conducted.

Postoperative adjuvant chemotherapy with taxol and carboplatin every three weeks, which is similar to that used for ovarian carcinoma, is normally initiated (Katsumata & Noriyuki, 2013). Our patient was given a total of six cycles of carboplatin and taxol. She was under surveillance for three years while the disease was under remission.

The majority of patients who present with advanced epithelial cancers of the fallopian tube will relapse with a median time to recurrence of sixteen months (Berek et al., 2021). The treatment for relapse fallopian tube

cancer is the same as relapse ovarian cancer. Parmar et al., (2003) proved that patients with a treatment-free interval of more than six months are considered to be platinum sensitive and commonly treated with platinum-based chemotherapy. Genetic testing and hormonal suppression should be considered in serous fallopian tube cancer cases since this treatment option has been applied in ovarian cancer cases (Simpkins et al., 2013). The rationale of hormonal suppression is to stop production of oestrogen from the ovaries or to reduce circulating oestrogen levels. This option has been well used as treatment for breast cancer and ovarian cancer. Hormonal suppression can be either hormone or hormone blocking drugs, for example a) GnRH agonists (example goserelin, leuprolide) b) oestrogen receptor blockade (example tamoxifen) c) oestrogen synthesis suppression, aromatase inhibitors (example letrozole, anastrozole, exemestane) d) oestrogen receptor downregulation, oestrogen receptor antagonist (example fulvestrant) e) androgen receptor blockade, antiandrogen (example flutamide) f) progesterone receptor blockade, progesterone receptor antagonist (example mifepristone, medroxyprogesterone, megestrol acetate) (Li et al., 2021). Aromatase inhibitors can be used to treat low grade serous ovarian cancer. Genetic testing and hormonal suppression might be the treatment option for our patient however, both were not given as an option. In view of significant change in practice over the last 20 years, patients have been routinely followed up with regular CA-125 testing after completion of chemotherapy. Our patient had a relapse after three years. She was again given platinum-based chemotherapy for the recurrence and has been under regular monitoring of CA-125 level and CT TAP yearly. Currently, she is on remission and coping well.

CONCLUSION

- Fallopian tube malignancy is a rare gynaecological tumour which occurs commonly in post-menopausal elderly women.
- It is not uncommon for fallopian tube malignancy to occur in young patients.
- Common symptoms are lower abdominal pain, abnormal watery vaginal discharge and a palpable pelvic mass.
- The management of fallopian tube malignancy are surgery and chemotherapy.
- Hormonal suppression should be considered in fallopian tube malignancy which occur in young patients.

CONFLICT OF INTEREST

The authors do not have any conflict of interest.

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