

Primary fallopian tube carcinoma presenting with a solitary metastasis in a Spigelian hernia

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Abstract

Primary fallopian tube carcinoma (PFTC) is a rare gynaecological tumour.

Histologically, this type of tumour resembles other tumours of the female reproductive tract, predominantly serous tumours. These cancers are often difficult to distinguish from ovarian cancers and are generally treated with the same standards of surgery and chemotherapy. Diagnosis of PFTC is rarely made preoperatively. Very often, the diagnosis is made either at the operating table or by the pathologist. In this report we present a rare case of PFTC with a solitary metastatic deposit in a right Spigelian hernia, the latter being the initial presenting complaint.

Key words

Fallopian tube neoplasm, diagnosis, tumour markers, Spigelian hernia

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Case report

A 65 year old nulliparous female presented at the surgical out-patient department with a two month history of a lump on the right side of her abdomen. The patient complained of some degree of abdominal discomfort especially when the lump became more prominent. Clinical examination revealed a partially reducible lump with a palpable cough impulse along the right linea semilunaris just below the level of the umbilicus. A diagnosis of Spigelian hernia was put forth. In order to confirm the diagnosis, a CT scan of the abdomen (Figure 1) was performed and this confirmed the presence of the Spigelian hernia. Following this a standard repair of the hernia was scheduled.

At operation the hernia was approached via a transverse skin incision over the lump. The anterior rectus sheath and external oblique aponeurosis were incised to expose the sac. Once the sac was opened, an encysted gelatinous mass was exposed. The cystic mass had a long pedicle extending down into the pelvis. The pedicle was ligated and excised. The cyst was sent *in toto* for histology.

The pathology report stated that the cystic mass measured 4.5x3.0x2.5cm. On sectioning, it was cystic, multilobulated, and contained yellowish, mucinous fluid. The cyst was smooth and contained a 0.7cm nodule. Microscopically (Figure 2), it was reported as a poorly differentiated adenocarcinoma in a peritoneal hernial sac. This could represent a primary peritoneal tumour or metastasis from other sites such as the ovary, however other possible primary sites had to be excluded. Therefore a CT scan of the abdomen and pelvis (Figure 3) was performed. This showed a slightly enlarged and irregular right adnexa. A serum CA-125 radioimmunoassay was performed and this was grossly elevated at 156.00 U/ml (1.9-16.3 U/ml). The case was discussed and a joint laparotomy with a gynaecologist was performed.

At operation the CT findings of an enlarged and irregular right adnexa was confirmed. Both ovaries, the left fallopian tube, omentum and pelvic and para-aortic lymph nodes looked normal and there was no evidence of intraperitoneal spread. Consequently, the gynaecologist proceeded to perform a total abdominal hysterectomy, bilateral salpingo-oophorectomy and omentectomy followed by a thorough peritoneal lavage.

The histology was that of an invasive poorly differentiated serous adenocarcinoma of the right fallopian tube which was ruptured and adhesions were extending between the tube and the attached right ovary. The latter was otherwise normal. The



Figure 1: CT scan of the abdomen depicting the right Spigelian hernia

findings were consistent with a primary fallopian tube carcinoma (Figure 4). The omentum was free of any metastatic deposit. The uterus and left adnexa were normal.

The patient made an uneventful recovery and was referred to the oncologist. She underwent 4 cycles of paclitaxel and carboplatin. Before her first cycle the CA-125 level was 8 U/ml. After the fourth cycle the CA-125 was 7.6 U/ml. One and three years later the CA-125 was still within the normal range. Her liver function tests and other tumour markers were and remained within normal range too.

Discussion

Primary fallopian tube carcinoma (PFTC) is a rare form of gynaecological tumour, with an incidence varying from 0.3% to 1.1% of all gynaecological cancers.^{4,43} Tubal carcinoma has been reported in females from 14 to 87 years of age, with a peak incidence in the early 60's.^{4,5} In fact our patient was in her mid-sixties. PFTC is more common in nulliparous females as was our case. This was established from a meta-analysis of cases reported between 1973 and 1992. This showed a nulliparity rate of 27.5% and a mean parity of 1.7%.⁵ Historically, Renauld reported the first case in 1847.⁶ Up to 1998 a total of 1500 cases

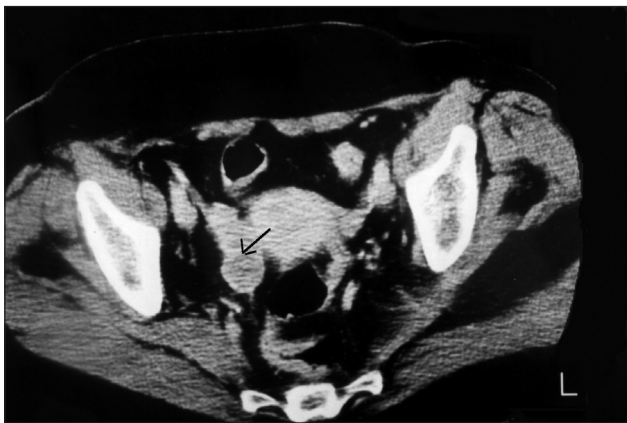


Figure 3: CT scan of the pelvis showing the enlarged and irregular right adnexa

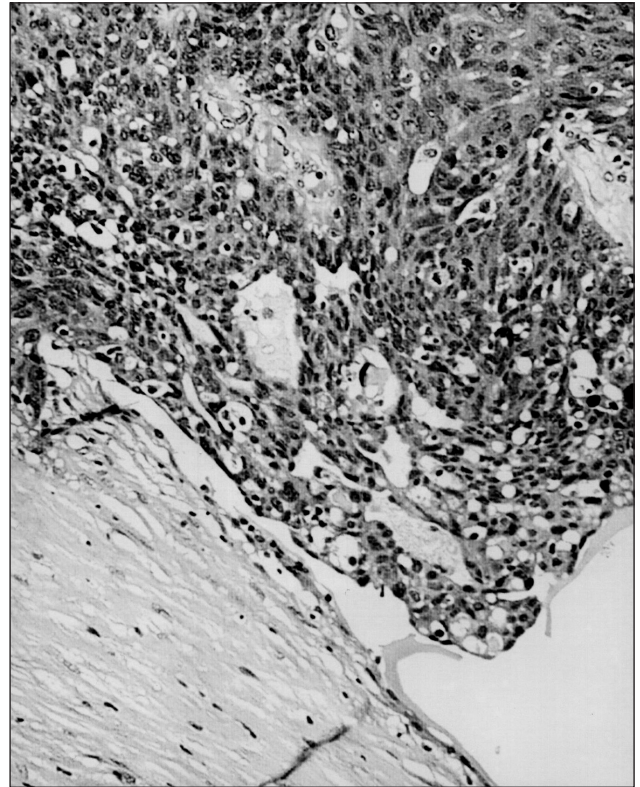


Figure 2: Histological view of the nodule in the peritoneal hernial sac - poorly differentiated adenocarcinoma



Figure 4: Poorly differentiated serous adenocarcinoma of the right fallopian tube

of tubal carcinoma have been reported either in small series or individual case reports.^{7,10} Major risk factors for fallopian tube carcinoma appears to be similar to those for ovarian carcinoma, oral contraceptives and childbearing both being strongly protective.¹⁵

The problem with these primary fallopian tube tumours is their difficulty in being diagnosed. The trio of abdominal pain, watery vaginal discharge, and metrorrhagia is considered by many as pathognomonic.⁶ Unfortunately, however, only a minority of cases present in this way. In fact, one of the papers reviewed, reported that 43% of patients were asymptomatic.⁵ In this specific paper by Nordan (1994), which dealt with a retrospective analysis of seven patients with PFTC, not one of these patients had a correct pre-operative diagnosis. Similarly, Butler *et al.* (1999) reported that in their series of 26 patients, no patient was correctly diagnosed pre-operatively.¹⁴ Figliolini *et al.*, in 1997 reported that over the previous 40 years the pre-operative diagnosis had actually increased from 0.26% - 6.38%.⁵ In the symptomatic group of patients preoperative diagnosis can be achieved by a combination of an abnormal cervical, elevated CA-125, ultrasonography or CT scan of the pelvis and laparoscopy. A study by Kujak *et al.* published in 2005 showed that a three dimensional ultrasound allows a precise depiction of tubal wall irregularities. Study of the vascular architecture in cases of fallopian tube malignancies could be further enhanced by 3D power Doppler imaging.¹⁶ The tumour marker Serum Cancer Antigen 125 (CA 125) is often elevated as was the case in our patient. In a reported series of patients with elevated CA-125, the ratio of epithelial ovarian to tubal cancer was 6:1.¹¹ A high serum CA-125 does not therefore distinguish primary ovarian from primary fallopian tube carcinoma. Besides, it is important to emphasize that CA-125 assays can be elevated in the absence of carcinoma and in the presence of other non related cancers. Therefore reliable pre-operative diagnosis requires a high index of suspicion and careful exclusion of alternative pathologies by radiological means. However, as in ovarian cancers, serum CA-125 are important markers of prognosis and therapeutic response to treatment independent of tumour stage. The pre-treatment CA-125 levels correlate well with the tumour stage but not with lymph node involvement, patient age or histological grade. This study also showed that in 90% of patients, a rise in the level of CA-125 preceded the clinical or radiological recurrence of disease by an average of three months.¹²

Several studies have cited that fallopian tube cancers have similar somatic genomic alterations compared with serous ovarian cancers and hence PFTC may arise in association with germ-line mutations of BRCA1 and BRCA2.¹⁷ The cumulative lifetime risk of developing hereditary breast ovarian cancer syndrome approaches 100%.¹⁸ These hereditary breast ovarian syndromes include fallopian and primary peritoneal cancers.^{19,20}

Management of PFTC depends on the stage of the disease, this being based on the FIGO system. Stage I and II patients

are known to do very well with primary surgical and adjuvant therapy, while the prognosis of Stage III/IV patients is much worse. Significant levels of long term survival can be achieved with aggressive treatment in the latter group.¹⁴ The surgical management includes 1) total abdominal hysterectomy and bilateral salpingo-oophorectomy, 2) omentectomy and peritoneal lavage and 3) selective para-aortic and pelvic lymphadenectomy. Adjuvant chemotherapy administration depends on the stage of the disease. The case we present was stage III and she underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy and omentectomy followed by adjuvant chemotherapy. Lymphadenectomy was not performed as there were no enlarged lymph nodes at laparotomy.

An extensive literature search regarding metastatic presentation of PFTC has revealed that apart from splenic metastases⁸ and inguinal lymph node metastases⁹ there has been no report of an isolated metastatic deposit from a PFTC in a Spigelian hernia.

The incidence of Spigelian hernia is not generally known but in one series of 1800 anterior abdominal wall herniae, an incidence of 0.5% was reported.¹ It is slightly more common in females and the median age is 50 years. The hernia is often impalpable, especially if the patient is obese and the only symptoms may be of vague abdominal pain. If a mass is present it may have a cough impulse and it may be reducible. However, 20% present with strangulation and intestinal obstruction.² It is not uncommon for its contents to be irreducible. Ultrasound and CT Scanning can easily confirm a Spigelian hernia and differentiate it from other abdominal wall masses such as abscesses, tumours, lipomas or haematomas.³ An online literature search revealed a report of a primary serous papillary carcinoma in a Spigelian hernia.²¹

Conclusion

This case report highlights the fact that an ordinary presentation such as a partially reducible hernia, as was the case, may belie a more sinister pathology. This illustrates the importance of submitting all specimens for histological analysis.

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