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Primary fallopian tube carcinoma: A case report and literature review

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Abstract
Primary fallopian tube carcinoma is rare and accounts for about 0.3% of all gynaecolgical cancers. Fewer than 2000 cases have been reported in literature. In routine practice, a case of tubal carcinoma is usually diagnosed as an adenxal mass, more commonly as an ovarian tumour. It is usually an intraoperative finding or a histopathological diagnosis.
A 45 year old postmenopausal woman, presented with the complaint of pain abdomen since 3 months. On per vaginal examination her uterus was retroverted, 10 to 12 weeks in size and a hard, fixed, nontender, irregular mass of 8 cm x 8 cm, in continuation with the uterus was felt in the left fornix.
The clinical and radiological findings with an elevated CA 125 suggested an ovarian tumour with a fibroid uterus. An exploratory laprotomy was done and in the left fallopian tube an exophytic growth 10 cm x 10 cm adherent to the lateral pelvic wall was detected. The left ovary was seen seperate from the mass. Histopathology report confirmed it as a papillary serous adenocarcinoma of the fallopian tube.

Keywords: primary fallopian, carcinoma, vaginal

Introduction
Primary fallopian tube carcinoma (PFTC) is a rare gynaecological malignant tumour and accounts for approximately 0.14 to 0.18% of all genital tract malignancies [1, 2]. In 1847, Renaud described fallopian tube malignancy and in 1888, Orthomann presented the first case report [5]. Only 2000 such cases have been reported so far in literature [1, 2]. The tumour must be located macroscopically within the fallopian tube or its fimbriated end to be considered a primary fallopian tube carcinoma [3].
Its incidence is estimated to be 2.9/1,000,000 to 5.7/1,000,000 [1, 5]. The etiology is unknown but it is associated with chronic tubal inflammation, infertility, tuberculous salphingitis and tubal endometriosis [1, 3]. Fallopian tube carcinomas have an inherent similarilty to ovarian cancers. Genetically, BRCA germline mutations and TP53 mutations association have been observed in both tumours [1].
It is seen to arise in post menopausal women. The clinical signs and symptoms are non specific and include lower abdominal, pelvic pain, serosangunious vaginal discharge and pelvic mass [1, 2, 3]. The rate of perioperative diagnosis was in the range of 0% to 10% and in most cases it is an intraoperative finding or a histopathological diagnosis [1, 4].
In comparison to ovarian carcinoma, it more often presents at an earlier stage but with a poorer prognosis but the principal of management is identical. We are reporting a rare case of PFTC in a 45 year old postmenopausal woman which was an incidental finding in staging laprotomy.

Case report
A 45 year old postmenopausal woman, para four, living four presented with the chief complain of pain abdomen since three months. The pain was insidious in onset, dull aching, located in the lower abdomen and not associated with any reliving or aggravating factors and was non radiating. On the admission day her, blood pressure was 130/90 mmHg, the pulse rate was 80 beats per minute, and her temperature was 37 °c. She was conservatively managed at various hospitals but her symptoms had not improved. Her haemogram, hepatic, renal function as well as blood sugar, urine examination were within normal limits.
On physical examination tenderness was noticed in the left lateral quadrant. The speculum examination showed a healthy cervix and vagina. On per vaginal examination her uterus was retroverted, 10 to 12 weeks in size and had a hard, mobile, irregular, nontender mass, of 8*8 cm, in continuation with the uterus which was felt in the left fornix. A soft cystic mobile mass 6*6 cm nontender was felt in the right fornix.
A transabdominal sonography showed that the borders of the uterus and the left ovary were not separately seen. In the same region a solid lobuated mass encasing the uterus was identified. multiple fibroids were seen arising from the uterus. the right ovary was visualized and appeared normal.
The MRI pelvis showed a 66*78.6 mm sized predominantly solid lesion in the left ovary with few cystic areas and multiple septations. The lesion appears homogenously hyperintense on T2, iso intermediate signal intensity on T1 showing homogenous enhancement on post contrast study. A 74 * 44 mm cystic lesion with few internal septations were noted in the right ovary. These features were suggestive of bilateral ovarian neoplasm. A well-defined round lesion measuring 28*29mm was noted in the posterior myometrial wall which appeared homogenously intense on both T1 and T2 weighted images suggestive of fibroids. The CA125 antigen was 370.53 U/ml. On the basis of the clinical and imaging results, bilateral ovarian neoplasms were suspected and a decision for staging

laprotomy was taken.

In the staging laprotomy, there was no ascites. A peritoneal wash with 100 ml NS was given and the fluid collected for sent for cyotological evaluation. The uterus was enlarged and 2 to 3 fibroids were seen in the posterior wall and in the anterior wall near the fundus. A left sided exophytic growth of 10 *10 cm with a ruptured capsule was seen arising from the fallopian tube. The growth was adherent to the lateral pelvic wall and posteriorly to the rectum, but separable from the descending colon. A 6 *7 cm unilocular paraovarian cyst arising from the fimbrial end was found and secondaries were seen over the omentum and the small intestine. Both the right and the left ovaries were normal.

A total abdominal hysterectomy with the bilateral salpingo-oopherectomy was done. She was staged as FIGO stage IIIB.

The frozen section report showed malignant surface epithelial tumour suggestive of papillary serous adenocarcinoma. The peritoneal wash fluid showed malignant cells, also consistent with metastatic adenocarcinoma. She was referred to a higher center for chemotherapy.

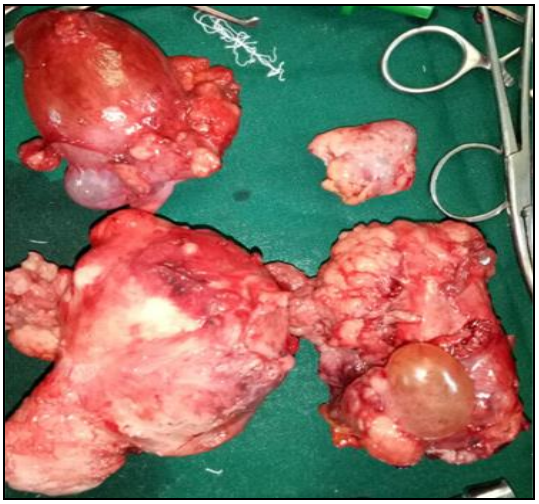

Fig 1: left sided fallopian tube mass with a right side paraovarian cyst

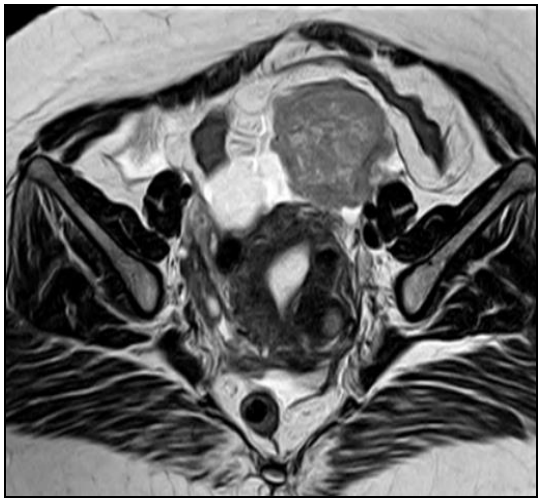

Fig 2: MRI T1 post contrast

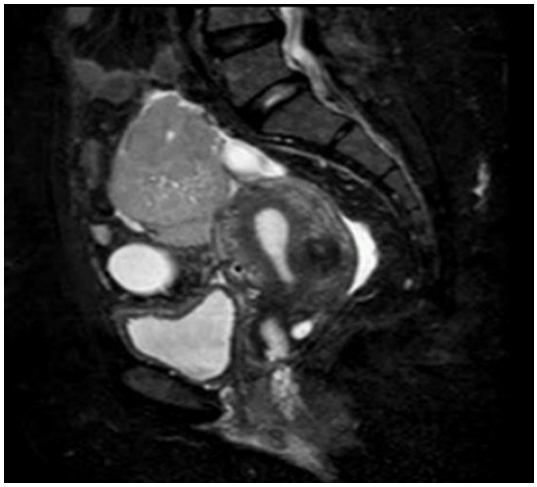

Fig 3: MRI Stir Image

Discussion

Primary fallopian tube carcinoma (PFTC) is a malignancy of the tubal epithelium and is one of the rarest carcinoma and are mostly adenocarcinoma on histological findings [1, 2, 3]. It accounts for less than 1% of all gynaecological malignancies and are mostly adenocarcinomas [1, 2]. It resembles epithelial ovarian carcinoma, but diagnosed at an earlier stage than the latter because of abdominal pain from tubal distension and a shorter history of symptoms.

The peak incidence is between 60 and 64 years of age with a mean incidence of 55 years [1, 2]. Few cases have been reported in adolescence; the most documented one being by Gatto in a 16 year old [2, 6]. The etiology of this cancer is unknown. There are no predisposing factors but it has been found to be associated with pelvic inflammatory disease as well as nulliparity, and subfertility [2]. PFTC has been described in high risk breast-ovarian cancer families with germ line BRCA1 and BRCA 2 mutations. Some studies have suggested that the frequency and structure of the chromosomal changes observed [BRCA-1 or BRCA-2 mutations] seen in PFTC had similarities with thoese found in breast, serous ovarian and uterine carcinomas and a common pathogenesis was claimed [2, 7].

Though our patient was postmenopausal and presented with a relatively short duration of symptoms, she did not have any predisposing factor and did not fall in the high incidence age group.

The patients present with a palpable pelvic / abdominal mass (61%), abnormal vaginal bleeding (47.5%), lower abdominal pain (39%) and an abnormal vaginal discharge (20%) [1, 2, 3]. The lower abdominal pain is colicky is a result of forced tubal peristalsis or dull as a result of tubal distension [1]. The Latzko's triad typical symptoms consists of intermittent profuse serosanguinous vaginal bleeding, colicky pain relived by discharge and an abdominal or pelvic mass was reported in 15% of the cases [1]. In our case, apart from dull abdominal pain, all of these symptoms were absent.

A pap smear positivity occurs in case of 10% to 36% [1]. A diagnostic curettage and pap smear was negative in our patient and the CA125 level was raised. The CA 125 is a useful marker for the diagnosis, assessment of response to treatment and the tumour recurrence [1, 2, 3].

The diagnostic criteria for PFTC was first established by Hu and colleagues and later modified by Slides. Accordingly, PFTC can be distinguished from other gynaecologcial malignancies (1) the main tumour arises from the endosalphnix, (2) the histological pattern reproduces the epithelium of the tubal mucosa (3) the transitition from benign to maliganant is demonstrable and (4) the ovaries and the endometrium are normal or contain tumour that is smaller than the tumour in the tube [1, 2, 3].

Imaging rountinely carried out for any suspicious malignancy of the genital tract carcinoma include ultrasound, computed tomography, and MRI. PFTC is difficult to diagnose radiologicaly and usually diagnosed as an ovarian neoplasm.

Table 1: Frequency of different histological subtypes of primary fallopian tube carcinoma

Histological Type	%
Serous	45-90
Endometriod	8-50
Mixed	4-20
Undifferentiated	7-12
Clear Cell	1.9
Transitional	12
Mucinous	3-8

All the carcinoma subtypes of ovarian cancers have been identified in the fallopian tube. 80 % of the tumours are serous with palliary, solid, glandular and micropalliary architecture. The second histological subtype is endometriod carcinoma with cribriform or solid areas of squamous or mucinous metaplasia. The other types are clear cell, mucinous, transitional or undifferentiated carcinomas [1, 2, 3] [table 1].

Transvaginal and transabdominal Ultrasound is an essential imaging technique in the diagnostic workup [1, 2, 3, 4]. The appearance of PFTC is usually based on the dominant component of the tumour – solid tumour or the hydrosalpinx, which maybe altered by serial imaging, reflecting the change in the amount of serous fluid in the tube. The former appears as a sausage shaped adnexal mass, and the latter appears as a fluid filled tubular adnexal structure, containing nodular or papillary solid components, or a multilocular cyst with a cog wheel appearance [1, 3].

On a CT scan, the lesion has attenuation equal to that of other pelvic soft tissue but the enhancement is less than the myometrium. MRI is a better diagnostic tool. On T1 – weighted MRI, the tumour is usually hypointense; and on T2-weighted MRI the tumour is homogenously hyperintense. Gandolium enchanced MRI most often detects solid, cystic components with papillary projections. The tumour infiltration of the bladder, vagina, pelvic side walls, pelvic fat, rectum is remarkably better with MRI than with CT or ultrasound [1, 3].

PFTC is richly permeated with the lymphatic channels which drain in the paraaortic lymphnodes through infundibulopelvic lymphatic's. It spreads by local invasion, transluminal migration, bloodstream and via the lymphatic's. The principal method of spread is the trans coelomic exfoliation of the cells throughout the pelvic and peritoneal cavity [2, 3].

The staging is based on Figo Staging system (Table 2) which requires a complete surgical approach. The stage of the disease is the most important prognostic factor [1, 2, 3, 4]. PFTC five year survival rates are about 68 to 76% for stage I, 27-42% for stage II and 0 to 6% for stage III and IV [1, 2, 3].

In our case, surgery was done, the tumour was histopathologically confirmed as palliary serous adenocarcinoma and a diagnosis of stage IIIb was established.

Table 2: Staging of primary fallopian tube carcinoma (PFTC)

FIGO		TNM
0	Primary tumor cannot be assessed	T_x
	No evidence of primary tumor	TO
	Carcinoma situ (pre-invasive carcinoma)	Tis
1	Carcinoma confined to fallopian tubes	T1
1(A)	Tumor confined to 1 tube without infiltrating the serosal surface: no ascites	T1a
1(B)	Tumor confined to both tubes without infiltering the seasonal surface: no ascites	T1b
1(C)	Tubmor confined 1 or both tubes with extension onto/through the tubal serosa or with positive malignant cells in the asceteis or positive peritoneal washing	T1c
2.	Tumor involving both tubes with pelvic extension	T2
2(A)	Extension and/or metastases to uterus and/or ovaries	T2a
2(B)	Extension to other pelvic organs	T2b
2(C)	Stage 2(A) or 2(B) with positive malignant cells in the ascites or positive peritoneal washing	T2c
3	Tumor involving 1 or both tubes with peritoneal implants outside the pelvis and/or positive regional lymph nodes	T3 and/or N1
3(A)	Microscopies peritoneal metastases outside the pelvis	T3a
3(B)	Macroscopic peritoneal metastases outside the pelvis>2 cm in greatest dimension	T3b and/or N1
3(C)	Peritoneal metastases more than>2 cm in greatest dimension and/or positive regional lymph nodes	T3c and/or N1
3(D)	Distant metastases beyond the peritoneal cavity. Positive pleural cytology and/or parenchymal liver metastases	M1

Abbreviation: FIGO, International Federation of Gynecology and Obstetries; TNM, Tumor-Nodes-Metastasis.

Surgery is the treatment of choice for PFTC and is similar to that of ovarian carcinoma's. The cytoreductive surgical technique involves the maximal removal of the tumour. The procedure of choice is bilateral salpingo-oopherectomy, omentectomy, selective paraaortic and pelvic lymphadenectomy for any stage of the disease. Postoperatively, a platinum based combination chemotherapy is used and the role of radiotherapy is less clear [1, 2, 3, 4].

In conclusion, the primary fallopian tube carcinoma is an extremely rare gynaecological malignancy. Postmenopausal

women are more susceptible. It has a late clinical presentation with a variety of non specific signs and symptoms and the pathogonomic symptom complex of "hydrops tube profluence "is rarely encountered. On imaging it is usually described as abdominal, pelvic or adnexal mass and is often confused with ovarian malignancies. It should be considered as a differential diagnosis while planning a surgery in pre and postmenopausal woman with unexplained uterine bleeding, pelvic pain, adnexal mass, abnormal cervical smear and complicated pelvic disease. Complete surgical resection or cytoreductive surgery with minimal residual tumour followed by combination adjuvant chemotherapy similar to that ovarian carcinoma is the mainstay of the treatment.

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