Original Article

A clinical cancer series of rare gynaecological malignancies

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Abstract

Angiomyofibroblastoma (AMF) is a rare, benign, soft-tissue tumor, which predominantly occurs in the vulvovaginalregion of middle-aged women. It is clinically important to distinguish an AMF from other stromal cell lesions. Here, we report the case of a 35-year-old woman with a rare, giant pelvic AMF, which showed a benign clinicalcourse. The Surface epithelial stromal tumours (adenocarcinomas), representing 80% to 90% of adult ovarian neoplasms, account for only 7% of malignancies in children. Here we are reporting a case of ovarian adenocarcinoma in a 14 years old female. PECOMA (Perivascular epitheloid cell tumor) is a rare mesenchymal tumour we are reporting a case of uterine PECOMA in a 45 years old female.

Keywords: AMF, Ovarian adenocarcinoma, PECOMA.

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CASE REPORT 1

A 35 years old woman presented with pelvic pain and intermittent dysuria since 6 months. Her medical history was unremarkable. Her menstrual cycle was regular, painless and she did not have dysmenorrhoea or menorrhagia. A physical examination didn't reveal any abnormalities. Her ultrasonography showed large well defined mixed echogenic mass measuring 11.2 X 10.4cm separate from posterior wall of uterus in Pouch of Douglas (POD) with peripheral vasularity. Computed tomography (CT) showed well defined mass lesion with moderately enhancing solid component sized 14 X 13.2 X 8.8 cm in pouch of douglas extending up to perineum arising from posterior vagina. No enlarged lymph nodes were seen in pelvic cavity. The patient underwent hysterectomy. A soft, regular shaped tumor was found behind the rectum. It had an intact capsule and did not adhere to or invade the peripheral tissues. Then complete local excision was carried out. The excised tumor was well circumscribed and measured 10 X 7 X 5cm. The cut surface was large flash like tissue. No haemorrhage, necrosis or cystic changes were observed. Microscopically tumor was well demarcated from surrounding fat tissue and was characterized by alternatinghyper cellular and hypocellular edematous zones containing thin walled blood vessels. Mitotic figures were absent. Interstitial myxoid degeneration was seen. Immunohistochemistry (IHS) revealed that tumor cells were positive for desmin, estrogen receptor (50 %), progesterone receptor (50%). The tumor was negative for smooth muscle actin, S – 100 Protein, CD 34, CD 117 and B – Catenin. These features indicated a diagnosis of AMF. Post operative USG was normal. Now patient is on closed follow up and long term disease free survival is under assessment.

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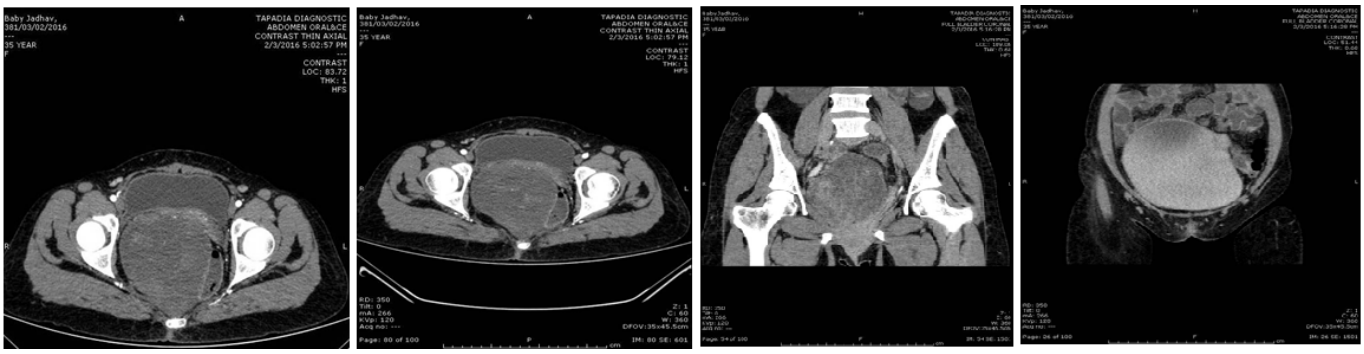


Figure 1: CT scan images for pelvic AMF

CASE REPORT 2

A 14 years old female presented with pain in abdomen and abdominal distension. Clinically, large mass of 32-34 weeks was palpable per abdomen with smooth surface and regular margins. USG abdomen andpelvis done on 19/04/2015 was suggestive of complex ovarian cyst of size 12 x 17cm with multiple loculi and septations. Serum CA 125 was 50 U/ml. patient undergone exploratory laprotomy with excision of ovarian mass on 20/04/2015. Histopathology report was suggestive of mucinous cystadenocarcinoma. Slides and blocks were reviewed as age is uncommon for ovarian epithelial malignancy. USG abdomen and pelvis on 04/07/2015 showed post operative status and rest was normal. Chemotherapy for 6 cycles in interval of 3 weeks was given from 30/08/2015 to 20/12/2015 (inj paclitaxel 250 mg and inj carboplatin 420 mg). patient tolerated chemotherapy well. CT scan abdomen and pelvis done on 09/01/2016 revealed right ovary bulky 4x2.6x3.8cm with heterogenous parenchymal enhancement and left ovary not visualized post operative status. CT scan chest revealed multiple well defined tiny soft tissue lesions with adjacent interstitial nodular thickening in apical segment of right upper lobe. Soft tissue nodule in superior segment of right lower lobe (multiple lung metastasis with adjacent lymphatic spread). Serum CA125, Serum AFP, Serum beta HCG and serum LDH were all within normal limits. Patient was asymptomatic for lung metastasis. PET-CT scan was done on 29/03/2016 which showed an ill defined hypodense lesion in right adnexa and was FDG non avid. No evidence of FDG avid pelvic or retroperitoneal lymphadenopathy. Tiny FDG non avid lung nodule was there on PET-CT which is of uncertain significance and needs follow up. No other FDG avid distant metastasis was there. Now patient is kept on close follow up. Long term disease free survival is under assessment.

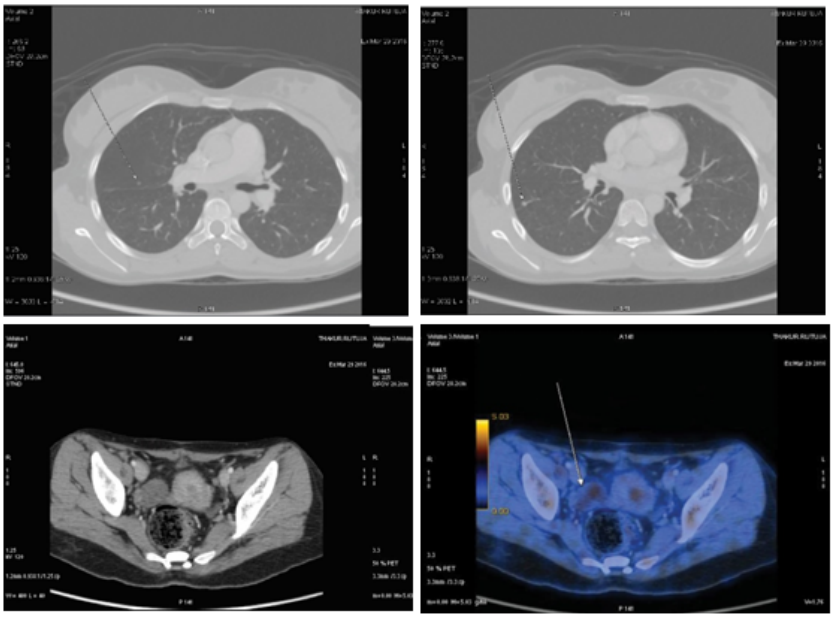
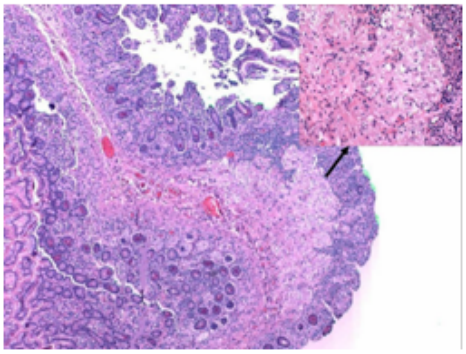


Figure 2: PET CT SCAN images showing tiny FDG non-Avid lung nodules and ill defined FDG non Avid hypodense lesion in right adnexa

CASE REPORT 3

Our patient was a 60years old female who presented with pain in abdomen, abdominal lump, dysmmenorrhea and menorrhagia since 6 months. Computed tomoghraphy revealed bulky uterus with large solid –cystic lesion of size 10x8cm in uterus with areas of necrosis with mild heterogenous enhancement suggestive of neoplastic etiology likely to be carcinoma endometrium. Then currettage was done which showed positive malignant cells. Patient underwent total hysterectomy with salphingo-oopherectomy with omentectomy with bilateral pelvic and para-aortic lymphadenectomy. Histopathological report revealed macroscopically an enlarged uterus with cervix 16x9x8cm.Endometrial cavity obliterated by fleshy grey pink necrotic looking tumor like mass of size 10x8x8cm.Myometrium thinned out, appears to be involved. Omentum and lymph nodes were free of tumor. The report was suggestive of poorly differentiated malignancy with differential diagnosis was low grade stromal sarcoma(NOS).IHC markers was done which revealed HMB-45 positive(40-45%), h-CALDESMON and MYOGENIN were negative suggestive of malignant PECOMA. After 3 months computed tomoghraphy was done which showed post-operative status with no abnormal enhancing soft tissue lesion in vaginal stump. Now patient is kept on closed follow up. Long term disease free survival is under assessment.



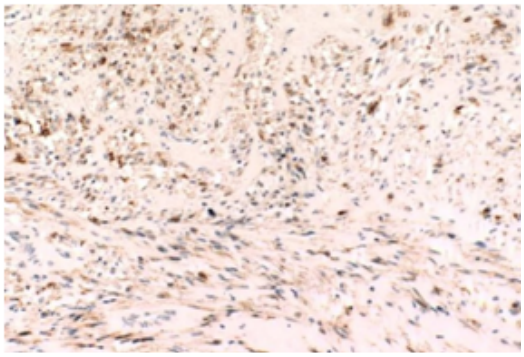


Figure 3: Tumour is composed of spindle to epitheliodcells with clear to eosinophilic cytoplasm and oval,hyperchromatic nuclei. Large bizare cells are seen.

Figure 4: Tumour is positive for HMB-45

DISCUSSION

CASE 1

AMF is a rare, benign, soft-tissue tumor that shows myofibroblastic differentiation and represents neoplastic proliferation of stromal cells. AMF was first describedby Fletcher *et al*. in 1992. AMF is composed of myofibroblastic cells and prominent thin-walled blood vesselswithin a fibromxyoid stroma. Its cause and pathogenesisare unknown at present. Since AMF is rare, no statistical data are available about its incidence in the general population. In most reports, AMF has occurred in women aged between 20 and 50 years, i.e., during the reproductive years. There are very rare reports of AMF occurring in malepatient. Most cases of AMF are benign, and only onecase with sarcomatous transformation has ever beenreported. A rare lipomatous variant of AMF with local invasion has also been reported. In most patients, AMF presents as a painless neoplasm located in the superficial regions of the lower female genital tract, such as the vulva and vagina[1], and aretherefore easily detected. Tumors arising in the cervix, uterus, urethral region sometimes present with obstructivesymptoms[2]. Tumors in the pelvis, iliac fossa or peritonealcavity are extremely rare, and usually grow insidiouslyand reach a massive size before they are detected. Several reports have analyzed the imaging features of AMFs. On perineal ultrasonography, AMFs appear as a soft-tissue mass with inhomogeneous mixed echogenicity, which corresponds to the cellular inhomogeneity found onhistopathological examination. Therefore, the ultrasonographic characteristics of AMFs may help to differentiate them from other mesenchymal neoplasms. On CT imaging, AMFs most likely show moderate-to-strongenhan cement, which may reflect the prominent vascularity of these tumors. In the current case, the small, hypoechoic area inside the mass observed on ultrasound examination may be attributable to the flesh-like structures seen on grossexamination.IHC showed that the tumor expressed estrogenand progesterone receptors, which suggests that it might have originated as a neoplastic proliferation of hormonally responsive mesenchymal cells. As AMF has a benign clinical course, it should be differentiated from other tumor-like lesions of the vulvovaginal region including Bartholin cysts, benignlipoma, fibroepithelial stromal polyps and cellularangiofibromas[1]. The treatment of choice for AMF is simple total excision, which is

usually curative, and there are almost noincidences of recurrences or metastasis after completeexcision, which confirms the benign nature of AMF. AMF shows no propensity for infiltrative growth.

CASE 2

Ovarian malignancies in children may represent an array of unique problems for the clinician who is more accustomed to diagnosing and treating ovarian neoplasm in adults. Although ovarian malignancies in children are rare (representing 0.2% of all ovarian neoplasms), their recognition and diagnosis are vital because they can be fulminant if treated inadequately. The average overall incidence of ovarian neoplasms is 1.7:100,000 per year [3].The rarity of ovarian tumors in children precludes a statistically significant compilation of the age related occurrence of specific tumors. The surface epithelial stromal tumour, representing 80-90% of adult ovarian neoplasms, account for only 7% of malignancies inchildren[4]. Endometrial and clear cell carcinoma are not found in the first two decades of life. The mucinous and serous cystadenocarcinomas are rare before puberty and have not been reported in children younger than 4 years old. The incidence of these coelomic epithelial lesions increases with advancing age. The gross and microscopic characteristics of thesetumours in adolescents are identical to those of similar neoplasms in adult. The tumors are bilateral in 10% of cases compared with 25% in adults. Initial symptoms may be absent or vague and the average delay between the clinical onset and histologic diagnosis is 3 to 4months. The most common presenting symptom is abdominal pain which is present in more than half of patients. Patients can also present with abdominal swelling with associated anorexia, weight loss, nausea or vomiting. Only 36% to 63% of cases are indentified correctly before surgery. A palpable ovary in the prepubertal patient is presumed to be abnormal because of the absence of gonadotropins. Although surgical resection and histologic examinations are the only definitive means of diagnosing an ovarian neoplasm, several radiographic, ultrasonographic, andlaboratory parameters can aid the clinician. Although malignant potential of surface epithelial stromal tumours in children (7.1 % to 13.5 % of cystadenomas) is less than is seen in adults, malignant neoplasm in children show a clinical course and mortality similar to what is seen in adults. Serous lesions have the poorest prognosis. Tumours of low malignant potential behave in a generally benign fashion, as if properly staged as IA, may be treated with salpingo- oopherectomy only. For malignant surface epithelial stromal tumors other than stage IA treatment includes total abdominal hysterectomy, bilateral salpingo-oopherectomy, omentectomy, pelvic / para-aortic lymph node sampling. Chemotherapy generally indicated for adjunctive therapy. Radiotherapy with a moving strip technique may be beneficial. The 10 year survival rate for all stages is 75 % primarily because of the preponderance of early stage lesions.

CASE 3

PEC was first described by Apitz in 1943 in terms of epithelioid feature in renal angiomyolipoma[5]. It is characterized by an epithelioid appearance with a clear to granular cytoplasm, round to oval, centrally located nuclei and a perivascular distribution.PEC can show any degree of atypia. PEComa is now a widely –accepted type of neoplasm. About 10% of reported uterine PEComas have been related to the tuberous sclerosis complex, an autosomal dominant disease characterized by mental retardation, seizures and the development of tumors in multiple organs. Diffential diagnosis of uterine PEComa include epitheliodleiomyosarcoma and endometrial stromal sarcoma[6]. The management of PEComas is variable. Surgical excision is the most common approach for malignant uterine PEComa, some patients have also undergone radiation and chemotherapy or even hormone therapy, but their effectiveness is still questionable[7].

CONCLUSION

CASE 1

Pelvic AMF is extremely rare but is benign. Its pre-operativediagnosis and differentiation from other soft-tissuetumors are challenging. The combination of radiological data, and histological and IHC findings can confirm the diagnosis.

CASE 2

Pediatric ovarian Adenocarcinoma is a rare entity. Serum CA-125 is used as a prognostic indicator as in adult ovarian malignancies. Surgical line of treatment and chemotherapy regimens are same in pediatric ovarian Adenocarcinoma as in adults. PET-CT scan should be considered as a follow up tool for disease and metastasis assessment. The aim of this study isto have an emphasis over such rare malignancies, so that proper treatment protocols could be followed and disease can be cured at an early stage.

CASE 3

We have reported here in the case of a uterine PEComa with malignant histologic picture. Immunohistochemical staining of HMB-45 should be performed on uterine epitheiloid cell tumor with clear or pale eosinophilic cytoplasm in order to avoid misdiagnosis. All uterine PEComas should be at least be regarded as tumors of uncertain malignant potential, and all patients diagnosed with uterine PEComa regardless of histologic features should receive long term follow up in order to determine the biological behavior of the tumor.

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