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Pendant on a Necklace – Case of Sertoli-Leydig cell tumour

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A 23 year-old lady was referred to our institution as a case of primary infertility lasting two years. She had had an irregular menstrual cycle since menarche (8/60-90) associated with dysmenorrhea. She had gained 15 kg in weight since her marriage. A systemic examination was unremarkable except for excessive fine hair all over the body and face. Abdominal and pelvic examinations were unremarkable. A routine vaginal scan showed a normal uterus, normal ovaries, and a hyperechoic mass of 4 x 4 cm near the left ovary with minimal free fluid in the pouch of Douglas. The differential diagnosis was a haemorrhagic or dermoid cyst of the left ovary. A routine infertility work-up showed raised testosterone of 4.2 nmol/l, but tumour markers were all normal. A magnetic resonance imaging (MRI) scan was requested as the mass was persistent after 3 months. The MRI revealed a normal uterus and ovaries with free fluid in the pouch of Douglas, a 4.2 x 3.2 x 4.1 cm heterogeneous solid mass with necrotic areas and intense contrast enhancement with high cellularity seen in the pouch of Douglas. There was no evidence of continuity of the mass or infiltration into the left ovary. There was no evidence of pelvic lymphadenopathy. The provisional MRI diagnosis was a malignant mass in the pouch of Douglas not infiltrating the ovaries. Further investigations for a possible primary source of malignancy were negative. A diagnostic laparoscopy revealed a pedunculated ovarian mass of 7 x 5 x 3 cm, dark in colour with solid and cystic components attached to the left ovary by a thick pedicle of 1 cm, a normal uterus and ovaries with ascitic fluid in the pouch of Douglas. The pedicle was ligated, cut and the mass was removed intact. Both ovaries were left in situ. The ascitic fluid was sent for cytology. The couple did not consent to removal of the affected ovary. The histopathology result for the mass was an intermediate to poorly differentiated Sertoli-Leydig cell tumor without heterogeneous elements. Peritoneal fluid was positive for malignant cells. The couple was counselled in detail about staging laparotomy and chemotherapy, but they refused. A testosterone and pelvic scan after 3 months were normal. Ovarian Sertoli-Leydig cell tumours are rare sex cord stromal tumour, accounting for less than 1% of ovarian tumors.[1] The majority of these tumors are benign, unilateral, only 3–5% being bilateral. These patients present with clinical features of virilisation due to excessive secretion of testosterone from the tumour, however 50% may have no endocrine symptoms.[2] The important prognostic factors for Sertoli-Leydig cell tumors are age of patient, degree of differentiation and stage of the tumour. Malignant potential as given by Young *et al.* is none in a well–differentiated tumour, about 11% in the intermediate type, 59% in a poorly differentiated tumour and about 19% in tumours with heterologous elements.[3] As these tumours are mostly unilateral, conservative treatment with unilateral salpingo-oophorectomy and evaluation of the contralateral ovary is recommended at reproductive age. For patients who have completed their family, hysterectomy and bilateral salpingo-oophorectomy is appropriate. For patients with poorly differentiated tumours, postoperative chemotherapy is recommended. In our case the histopathology of the tumour showed an intermediate to poorly differentiated Sertoli-Leydig cell, but the couple refused staging surgery or chemotherapy. She is still being followed up clinically.

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Abrupt onset of unilateral third nerve palsy secondary to central nervous system relapse in acute myeloid leukaemia

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Isolated central nervous system (CNS) relapse following allogeneic stem cell transplantation (allo-SCT) is rare and poses a major therapeutic obstacle to cure in acute myelogenous leukaemia (AML). CNS relapse in a leukaemic patient ranges widely from less than 10% to 30% and is associated with a high rate of treatment failure when it occurs following allo-SCT. Acute unilateral third nerve palsy is an unusual early manifestation of such an event. A 25 year-old male patient with a diagnosis of AML was referred to the Sultan Qaboos University Hospital Eye Clinic with a 3 days history of inability to move his left eye inwards. There was history of double vision associated with his problem. The patient was subjected to detailed ocular, medical and radiologic evaluations. Ophthalmic examination of the right eye was unremarkable. Examination of the left eye showed partial blepharoptosis of the upper eyelid and absent adduction and elevation of the eyeball. Depression was moderately reduced. Abduction was normal. The pupil was mildly dilated with absent direct and consensual reaction to light. The patient maintained an intact corneal reflex and normal looking optic disc. Neurological examination revealed no neurological deficits apart from above-mentioned features of left 3rd cranial nerve palsy. Four months prior to the presenting problem, the patient had had allo-SCT, but from that time there had been no evidence of CNS involvement. A magnetic resonance imaging (MRI) examination with intravenous gadolinium diethylenetriamine penta-acetic acid (Gd-DTPA) of the orbit and brain showed enlargement and enhancement of the cisternal and cavernous portions of the left third cranial nerve. There was no meningeal enhancement or parenchymal brain lesion. A lumbar puncture to evaluate the cerebrospinal fluid (CSF) showed the presence of myeloblasts. Based on the above findings, a diagnosis of CNS relapse was made and treatment with intrathecal methotrexate was initiated. Although most cranial nerves may be difficult to visualise with MRI scanning due to their small diameter and complex anatomic course, the oculomotor cranial nerves can be visualised and reliably assessed by standard MRI sequences, not only in the subarachnoid space and cavernous sinus but also in the orbit. Rapid developments in the MRI have enabled evaluation of the entire course of normal as well as abnormal cranial nerves. Indeed, imaging has been dramatically improved with the use of high resolution MRI. This case highlights the value of MRI in the early detection and accurate evaluation of cranial nerve lesions, thereby aiding in the clinical diagnosis, treatment and prognosis assessment of patients with CNS relapse in leukaemia.

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Successful Management of a Case of Gestational Trophoblastic Neoplasia in a Young Woman

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A 23 year-old, recently married Omani lady was admitted with 7 weeks of amenorrhoea, abdominal pain and bleeding per vagina. Her abdominal examination was unremarkable; a bimanual examination showed a uterine size of 12 weeks gestation with a closed cervix. Serum β hCG was 284, 263 IU on admission. An ultrasonogram revealed a "snowstorm pattern" diagnostic of a complete hydatidiform mole. Suction evacuation and curettage was performed after the initial work-up. A histopathological examination confirmed complete hydatidiform mole. Weekly follow-up showed rising β hCG 3 weeks after evacuation. Staging investigations like computerised tomography (CT) of the chest and magnetic resonance imaging (MRI) of the abdomen and pelvis showed two tiny deposits in the lung and a myometrial invasion of 2 cm size, confirming low risk gestational trophoblastic neoplasia. After counselling, single agent chemotherapy was started with methotrexate. Actinomycin D was added later due to poor response to treatment as evidenced by suboptimal β hCG regression. β hCG returned to normal in 10 weeks time and remains negative on follow-up. Gestational trophoblastic diseases (GTD) include a spectrum of diseases with wide range of biologic behaviour. It includes the benign form, hydatidiform mole; locally malignant form, invasive mole, and frankly malignant form, choriocarcinoma. Although persistent, gestational trophoblastic tumours develop most commonly after a molar pregnancy, they may follow any gestation. The incidence of hydatidiform moles varies from 0.1 to 1%; 8–15% of them develop malignant sequelae.[1] β hCG is the tumour marker used for the diagnosis and follow up of GTD. Hydatidiform moles need close follow-up due to their malignant potential. Recommended follow-up is weekly β hCG testing until 3 negative values, followed by monthly for another 6 months. Pregnancy should be avoided during this period. Invasive moles are characterised by local invasion into the myometrium with or without lung metastasis and present with persistently elevated β hCG, as in our case. Choriocarcinoma usually presents with symptoms of metastases like cough, dyspnoea, haemoptysis, headache, convulsion, stroke, and vaginal bleeding.[2] The World Health Organization (WHO) scoring system based on prognostic factors is used for the risk stratification of the disease. A score of 6 or less is low risk disease and is treated with single agent chemotherapy. A score of 7 or greater is high-risk disease, which needs multiagent chemotherapy with or without local radiation/surgery.[3] Choriocarcinoma is potentially curable and future fertility is unaffected if promptly treated. Women can conceive 1 year after completion of chemotherapy; there is no increased risk of pregnancy complications or congenital anomalies. The risk of recurrence of molar pregnancy is 1–3%, which should be ruled out by an early ultrasound.[4]

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This case was presented at the Sultan Qaboos University Clinical-Pathological Conference on 28th October 2009 with the title "Through the vine (vain) yard"

Pre-eclampsia or acute fatty liver of pregnancy

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We report the case of a 36 year-old gravida 3 para 2 woman who presented at 34 weeks gestation with malaise, nausea, vomiting and feeling unwell. She underwent an emergency caesarean section for non-reassuring foetal heart status. The postoperative course was complicated by severe postpartum haemorrhage and disseminated intravascular coagulation requiring multiple blood transfusions. The patient developed chronic hypoalbuminaemia and jaundice. A presumptive diagnosis of acute fatty liver was made and supportive care resulted in a good outcome. Acute fatty liver of pregnancy (AFLP) was first described by Sheehan in 1940.[1] It is a life threatening obstetrical emergency that can lead to high maternal and perinatal morbidity and mortality. The aetiology of the disease is unknown with an incidence of 1 in 10,000 to 1 in 15,000.[2] The most common presentation is malaise, nausea, vomiting and epigastric pain followed by jaundice. The laboratory findings are usually of mild to moderate increase in serum transaminases, hyperbilirubinaemia, hypoglycaemia and deranged coagulation profile.[3] Liver biopsy is the gold standard for diagnosing AFLP, but due to the presence of coagulation abnormalities, the diagnosis is usually made by clinical and laboratory findings.[1] Bleeding and disseminated intravascular coagulation (DIC) are one of most common complications of AFLP. Other complications include hepatic encephalopathy, sepsis, ascites, acute renal failure, respiratory distress syndrome and pancreatitis. Due to the high maternal and perinatal mortality, early diagnosis, prompt delivery and supportive care are required.

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This case was presented at the Sultan Qaboos University Clinical-Pathological Conference on 28th April 2010 with the title "The mysterious fountain"