

Asymptomatic Ovarian Mucinous Cystadenoma with a Solid Mural Leiomyoma

Case report and brief review

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الورم المخاطيني الغدي الكيسي عديم الأعراض في المبيض المصحوب بورم عضلي أملس جداري صلب

تقرير حالة مع مراجعة موجزة

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الملخص: ورم المبيض المخاطيني قد يتضمن عقد جدارية حميدة أو خبيثة. العقد الجدارية الحاوية على الورم العضلي الأملس هي علة حميدة نادرا ما تصاحب أورام المبيض المخاطيني. نعرض هنا حالة عقدة جدارية حاوية على ورم عضلي أملس ناشئة عن ورم مخاطيني غدي كيسي حميد لامرأة في التاسعة والعشرين من العمر تعاني من كتلة كبيرة غير متجانسة في البطن. بعد إجراءات تقييم الحالة ما قبل الجراحة، خضعت الحالة لعملية فتح البطن بسبب الشك في وجود ورم المبيض الخبيث. فحص النسيج المخبري أثبت الطبيعة الحميدة للعقدة الجدارية.

مفتاح الكلمات: ورم المبيض، ورم غدي كيسي مخاطيني، الورم العضلي الأملس الجداري، الكيمياء المناعية للأنسجة، تقرير حالة، عمان.

ABSTRACT: Mucinous neoplasms of the ovary may have associated benign or malignant mural nodules. A leiomyomatous mural nodule is a rare, benign lesion associated with mucinous tumors of the ovary. We report a case of a mural leiomyomatous nodule arising in a benign mucinous cystadenoma in a 29-year-old woman who presented with a large heterogenous abdominal mass. After pre-operative evaluation, exploratory laparotomy was performed upon suspicion of ovarian malignancy. A pathological examination confirmed the benign nature of the mural nodule.

Keywords: Ovarian tumor; Mucinous cystadenoma; Leiomyoma, mural; Immunohistochemistry; Case report; Oman.

CYSTIC TUMOURS OF THE OVARY, WHETHER benign or malignant, may be associated with mural nodules of various types. These mural nodules are composed of an epithelial or stromal component, or a mixture of both. Sarcoma, sarcoma-like mural nodules (SLMN), and anaplastic carcinomas in mucinous cystic tumours of the ovary have been well described.¹ However, a leiomyomatous mural nodule within a mucinous cystic ovarian tumour is a rare, benign, spindle cell lesion and there have been only a few reported cases. We describe a case of large benign mucinous cystadenoma with a leiomyomatous mural nodule that clinically and radiologically raised the possibility of a malignant ovarian tumour. The benign nature of the nodule was confirmed by a microscopic and

immunohistochemical examination.

Case Report

A 29-year-old unmarried woman with a large abdominal mass was referred to our hospital from a private institution. The abdominal mass was an incidental finding discovered when she sought medical advice for a 2 x 2 cm mobile mass in the right hypochondrium. She denied any recent weight loss or change in bowel habits. She had regular menstrual cycles with her last period three weeks prior to admission. There was no relevant medical or surgical history in the past. She had a family history of malignancy—an elder brother with a

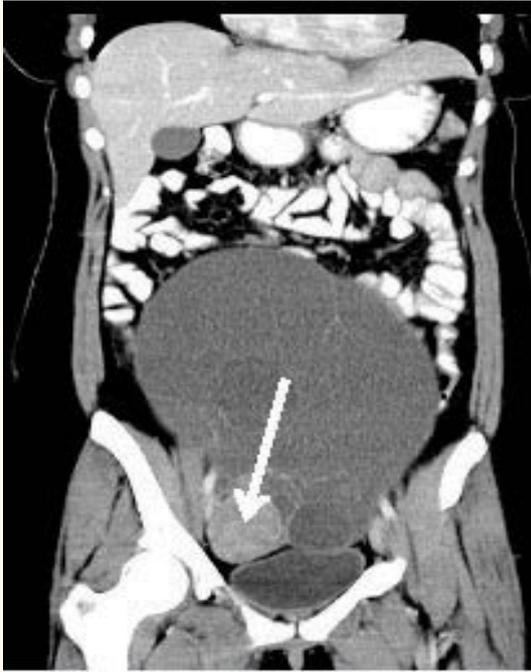


Figure 1: Computed tomography coronal re-formatted image with contrast showing an enhanced solid component.

brain tumour and an uncle with hepatic cancer.

On examination, her vital signs were normal with a body mass index (BMI) of 32. An abdominal examination revealed a smooth non-tender mobile mass arising from the pelvis corresponding to a uterine size distension of 28-weeks' pregnancy. A 2 x 2 cm lipoma was felt below the right costal margin. An abdominal ultrasonogram showed a large, predominantly cystic mass with multiple locules and a solid area of about 4 x 4 cm in the right lower region. Computed tomography (CT) and magnetic resonance imaging (MRI) scans revealed a 22 x 19 x 10 cm multiloculated cystic mass arising from the left ovary. There was a solid component measuring 5 x 4 x 4 cm with homogenous enhancement in the right lower part of the tumour, raising the possibility of malignancy [Figure 1]. The right ovary, uterus, and other abdominal organs looked normal with no ascites or lymphadenopathy. A chest X-ray was normal. The impression was ovarian cystadenoma/cystadenocarcinoma.

Blood investigations showed Hb 14 g/dl and a normal coagulation screen with normal renal and liver functions. Tumour markers such as cancer antigen (CA) 125, carcinoembryonic antigen (CEA), alpha-fetoprotein (AFP) and beta-human chorionic gonadotropin (β HCG) were normal. The patient



Figure 2: Gross specimen of ovarian cystic mass with mural nodule (arrow).

was scheduled for an exploratory laparotomy after arranging frozen sections. The laparotomy revealed a smooth-walled predominantly cystic mass of 22 x 18 x 10 cm in place of the left ovary with the left fallopian tube stretched over it. The uterus, right tube, and right ovary looked normal. There were no ascites, pelvic or para-aortic lymphadenopathy, or any other evidence of intra-abdominal metastasis. Peritoneal washings for cytology, and left salpingo-oophorectomy and an omental biopsy were performed. The postoperative period was uneventful.

The cystic ovarian mass submitted for frozen section weighed 2,400 grams and measured 22 x 18 x 8 cm. The surface of the cyst was intact with no papillary excretions; a solid whitish nodule measuring 4 x 3 x 3 cm was present on the wall of the cyst [Figure 2]. Sectioning revealed a multilocular cyst containing watery, non-haemorrhagic, mucinous fluid. There were small cystic spongy areas. The cyst wall thickness ranged from 0.2 to 0.4 cm. A cut section of the solid nodule on the wall of the cyst revealed a whitish whorled appearance. There were no haemorrhages or necroses in the cystic areas or in the solid nodule.

Haematoxylin and eosin (H & E) stained tissue sections from the cystic area showed large cystic areas lined by a single layered mucinous columnar epithelium with basally placed nuclei. Sections of the solid area revealed a lesion consisting of interlacing bundles of spindle cells with elongated nuclei and eosinophilic cytoplasm [Figure 3]. There was no nuclear pleomorphism and the mitosis was 2/10 HPF. The cystic and solid areas did not show any haemorrhages or necrosis.

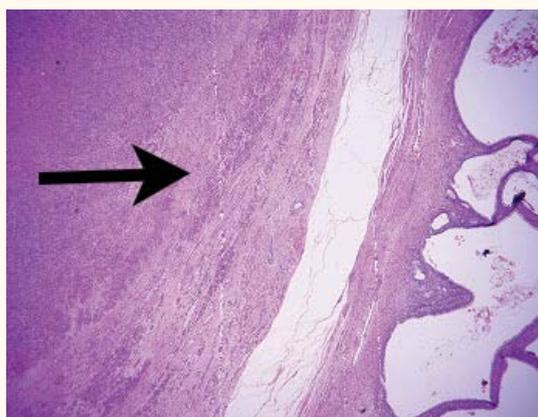


Figure 3: Haematoxylin & eosin stains. Magnification x 20 showing mural nodule (arrow).

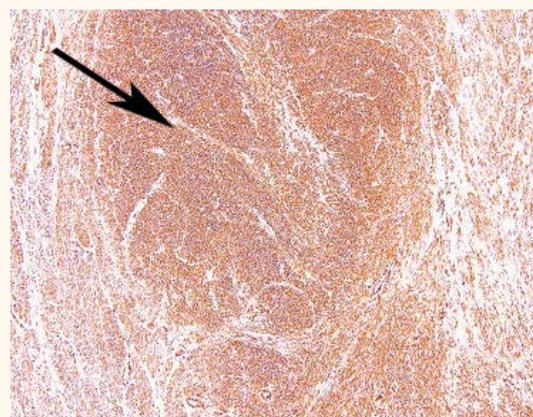


Figure 4: Spindle cells of mural nodule stained with smooth muscle actin x 10.

These spindle cells stained positive with smooth muscle actin (SMA) in immunohistochemistry [Figure 4]. They were negative for monoclonal antibodies anti-cytokeratin AE1/AE3 and CD117. A diagnosis of mucinous cyst adenoma with mural leiomyoma was made.

Discussion

The occurrence of mural nodules in serous or mucinous ovarian tumours is a rare but well-established entity. Mural nodules can develop in benign, borderline, or malignant ovarian tumours. They are a heterogeneous group of lesions and can be benign, reactive, or neoplastic in nature.² These mural nodules are seen as solid lesions on the wall of the tumour or project into the cyst lumen. Being a part of a large cystic ovarian tumour, they can mimic malignancy both clinically and radiologically. Reactive type mural nodules may mimic malignancy in histopathology as well. The benign and reactive mural nodules have a better prognosis; hence, it is important to differentiate them from the malignant mural nodules.^{2,3}

Reactive type mural nodules, also called sarcoma-like mural nodules (SLMN), occur in younger females and are usually sharply demarcated small lesions. There are three histological types of SLMNs: epulis-like, the pleomorphic and spindle cell type, and the histiocytic type. Malignant mural nodules are fibrosarcomas and anaplastic carcinomas; they tend to occur in older females. When a mural nodule is composed of spindle cells, it needs careful evaluation to assess whether it

represents a reactive spindle cell proliferation or a neoplastic process. Low-grade spindle cell lesions could be underdiagnosed as benign spindle cell proliferations.^{1,4-6} Immunohistochemistry will help to elucidate some of these diagnostic difficulties but awareness of the different types of mural nodules and a thorough assessment of morphology are still the key to the diagnosis.^{7,8} Pathogenesis of mural nodules in ovarian tumours is not clear. More case studies are needed before the pathogenesis of this rare entity can be fully understood.²

To the best of our knowledge, this is the fourth report of a leiomyomatous mural nodule in a benign mucinous cystadenoma in English medical literature, and the youngest patient reported so far; size-wise this is the second largest tumour. The first case was described by Lifschitz-Mercer *et al.* in 1990, where the presence of a leiomyomatous mural nodule was confirmed by immunohistochemistry.⁹ The second report was one of the five cases of cystic ovarian mucinous tumours with mural nodules reported by Nicholas *et al.*⁸ The third report of a mural leiomyomatous nodule in benign mucinous cystadenoma was published by Hameed *et al.* in 1997.¹⁰ In their study of five cystic ovarian mucinous tumors with spindle cell mural nodules, Nicholas *et al.* found that immunohistochemistry was useful in distinguishing the variant forms of spindle cell mural nodules in cystic ovarian mucinous tumours.⁸ They found that malignant spindle cell nodules co-expressed both cytokeratin and vimentin, whereas leiomyomatous nodules were negative for cytokeratin but positive for vimentin, desmin, and muscle-specific actin.^{7,8}

The prognosis of each patient depends on the

type of mural nodule associated with the tumour. Malignant mural nodules have been reported to have a poor prognosis despite receiving postoperative adjuvant therapy, with 50% mortality within 5 years.¹ Several case reports have shown a poor prognosis, depending upon the International Federation of Gynecology and Obstetrics (FIGO) staging associated with a sarcomatous/carcinomatous nodule, as some tumours may behave in an aggressive manner. De Rosa *et al.* stated that these tumours should be treated in the same way as high-grade conventional ovarian carcinoma despite a macroscopically favourable presentation.¹¹ In contrast, those with leiomyomatous nodules or sarcoma-like mural nodules have an excellent prognosis and their presence does not influence the prognosis of the ovarian tumour associated with it, provided they are well-demarcated and lack invasion to the surrounding tissue or vascular spaces. The existence of sarcomatous nodules combined with the SLMN is another rare entity that has been reported and necessitates a careful histologic analysis for treatment and the determination of prognosis.² Classification of these mural nodules is imperative in planning a patient's postoperative therapy and follow-up, and has a general bearing on the overall prognosis of the ovarian tumour it is associated with.^{11,12} Our patient did not require any postoperative treatment as the mural nodule was benign. On follow-up after 6 months, the patient was asymptomatic and a pelvic ultrasound was unremarkable.

One of the criteria which differentiates a malignant cystic ovarian mass from that of a benign cystic ovarian mass is the presence of a solid component. The larger the solid component in a cystic mass, the higher is the likelihood of malignancy. This was of important relevance to our case since the patient was unmarried and in the reproductive age group; hence, a more conservative treatment was desirable, which would have been difficult had this mass been malignant. The presence of the solid nodule in this mass did raise a suspicion of malignancy, although other factors like the unilaterality, the presence of a cystic mass in the younger age group, its asymptomatic nature despite it being a large mass, and the absence of ascites and normal tumour markers were all indicators of the benign nature of this tumour. In such a scenario, modalities like

a contrast CT/MRI, histopathological analysis, and immunohistochemistry can play a useful role in management, as has been demonstrated in this case.

Conclusion

Mucinous neoplasms of the ovary may be associated with benign or malignant mural nodules. These nodules appear as solid lesions radiologically, raising the suspicion of malignancy. Awareness of the heterogeneous nature of the mural nodules can help the clinician in planning the appropriate management of patients who have large cystic ovarian masses with solid nodules.

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