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RETROPERITONEAL MULLERIAN CYST: A CASE REPORT

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ABSTRACT

Mullerian cyst of the retroperitoneum is extremely rare. Only 15 cases have been reported in the literature. We report a case of a 41 year-old woman treated for 2 retroperitoneal mullerian cysts misdiagnosed as ovarian tumors that were completely resected surgically. With a short review of the literature, we focus on clinical, radiologic and histological features of this lesion.

Keywords: Mullerian cyst, Retroperitoneum, Surgery.

INTRODUCTION

Mullerian cyst of the retroperitoneum is extremely rare. It is characterized by the development of mullerian epithelium-lined cyst in the retroperitoneum similar to the lining epithelium of the fallopian tubes and the endometrium. We report a case of retroperitoneal mullerian cysts misdiagnosed as ovarian tumors, with a review of the literature focusing on clinical, radiologic and histological features of these lesions.

CASE REPORT

A 41 year-old woman, with no medical history, was referred to our unit for abdominal mass. The abdominal volume had increased gradually during the last 16 years. There were no genital, urinary or gastro-intestinal complaints. Abdominal palpation revealed a mass extending from the pelvis to the epigastric area. MRI showed two cystic masses with a low-intensity signal in a T1-weighted MRI image, and a high-intensity signal in T2-weighted MRI (Figure 1).

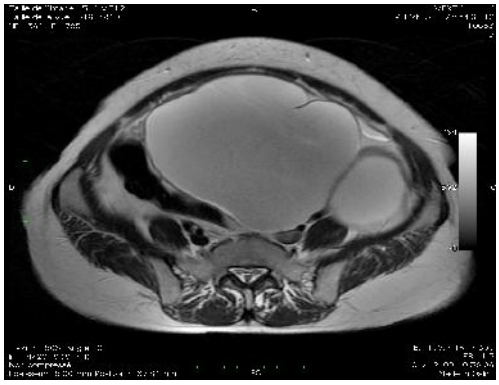

Figure 1: T2 weighted MRI showing the two cysts with high intensity signal

Within the cysts, there were thin septations that were enhanced after gadolinium injection. The biggest cyst contained haemorrhagic component. The cysts were occupying the pelvis and extending to the abdominal cavity, repressing laterally the intestine. The radiologist described them as bilateral ovarian masses. Serum CA-125 was within normal range.

The patient had a midline laparotomy. The ovaries were normal. The left colon was pushed medially by two retroperitoneal cystic masses. The first measured 20/15/5 cm and the second 25/35/20 cm and both had no connection with adjacent organs.

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The left kidney was pushed upward and the left ureter was identified medially to the cysts. We performed a complete removal without disruption of the cysts (Figure 2). The post-operative period was uneventful and the patient left the hospital five days after the procedure.

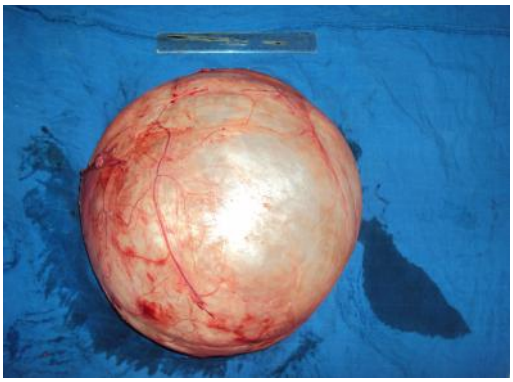

Figure 2: Surgical specimen mullerian cyst of 30 cm

Histology showed that the cysts' wall was composed of a dense acellular fibrous tissue. No other structures were found in the surrounding tissue. The epithelium lining the wall cysts consisted of a single layered ciliated and cuboidal serous cell containing round nuclei without any atypia (Figure 3). The features were consistent with serous cystadenoma of mullerian type.

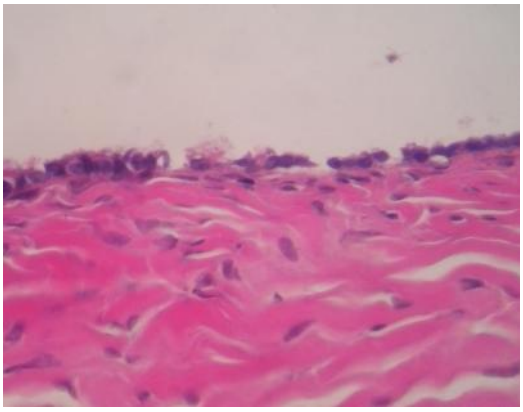

Figure 3: Microphotography of the cystic wall showing a single layered ciliated and cuboidal serous cell containing round nuclei without any atypia

DISCUSSION

Mullerian cyst of the retroperitoneum is rare, only 15 cases have been reported in the literature [1]. It is thought to be a subtype of urogenital cysts, the pathogenesis mechanism for the development of mullerian epithelium-lined cyst in the retroperitoneum is not clear. Three main theories are postulated. Retroperitoneal tissue may include aberrant embryologically-derived mullerian ducremnants that might have the capacity to grow in later life under the influence of abnormal hormonal stimuli. Alternatively, the coelomic epithelium or peritoneum may undergo differentiation to become serous/tubal-type epithelium, later invaginating into the underlying tissue and eventually losing its connection with the surface, producing a cystic structure. Some authors believe that ectopic endometrial tissue (endometriosis), as transplanted by retrograde menstruation or following pelvic surgical instrumentation, may give rise to retroperitoneal cysts [2]. In all reported cases, patients are adult women and age ranges from 38 to 80. The most common symptoms are abdominal discomfort and palpable abdominal mass. In two reported cases, patients were asymptomatic and the cysts were discovered incidentally [1]. At computed tomography, mullerian cyst manifests as a unilocular or multilocular thin-walled cyst containing clear fluid [3]. Magnetic resonance imaging is the best radiological method to determine the nature of the cystic lesion, such as serous or bloody but it was described in only one previously reported case of retroperitoneal mullerian cyst [4]. Retroperitoneal mullerian cyst has a low-intensity signal in a T1-weighted MRI image and a high-intensity signal with low-intensity septation in a T2-weighted MRI. In our case, this aspect was reported by the radiologist, but the mullerian cysts were misdiagnosed as ovarian tumors. The diagnosis of retroperitoneal mullerian cyst is difficult, pre-operatively, due to the lack of pathognomonic clinical and radiologic features. The diagnosis is based on the histological findings of ciliated columnar and endometrioid epithelium similar to the lining of the fallopian tubes and the endometrium [5]. Immuno-histochemical study of lining cells is positive for cytokeratins CK7, CK18, CAM 5.2, AE1/AE3, epithelial membrane antigen, CA-125, estrogen and progesterone. Staining for CK20, CEA, calretinin and CD10 is negative [6]. Unfortunately, in our case, immuno-histochemical study of lining cells was not performed.

Retroperitoneal mullerian cysts should be completely excised [7]; Optimally, with minimal disturbance of the surrounding normal structures [2]. In our case, cysts had no intimate contact with adjacent organs, and complete cysts excision without effraction was performed. External marsupialization, internal drainage or simple aspiration are associated with a high rate of morbidity and risk of recurrence and should be discouraged [2].

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