

Primary Retroperitoneal Mucinous Cystadenoma: A Rare Case Report

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Abstract: Primary retroperitoneal tumours of mucinous type are very rare and are further sub-divided into benign borderline and cystadenocarcinoma. Diagnosis of retroperitoneal tumours should not be delayed as the majority are malignant. We describe a case of 40 years old female who presented with complain of painless left side flank swelling from last 3 months. Her transvaginal ultrasonography revealed a large well marginated cystic lesion in supraumbilical region and left hypochondrium with CT abdomen further defining the lesion. Wide local excision of the cyst was done under GA. The histological diagnosis was confirmed as primary retroperitoneal mucinous cystadenoma (PRMC). In conclusion Primary retroperitoneal mucinous cystadenoma are rare, before diagnosis carefully search for ovary or other site and excision as early as possible because can be transformed in to malignancy.

Keywords: Primary, Retroperitoneal, Mucinous, Cystadenoma

INTRODUCTION

Retroperitoneal tumours account for less than 0.2% of all neoplasms [1]. Primary retroperitoneal tumours of mucinous type are very rare. They have the same macroscopic, morphologic and ultrastructural characteristics that of the ovarian mucinous processes [2]. The biological behavior and histogenesis of such tumors remain speculative because of the limited number of reported cases. The case we present was diagnosed with a primary retroperitoneal mucinous cystadenoma of benign type.

CASE REPORT

40 years old female with complains of painless abdominal swelling for 3 months which was gradually increasing in size. She did not have any significant past or medical history. On abdominal examination a non-tender 12 cm mass was palpable in the left lumbar region. Laboratory investigations revealed a normal full blood count, liver enzymes as well as urea and electrolytes.

Transvaginal ultrasonography demonstrated a 15x13x11cm well marginated cystic mass in supraumbilical region and left hypochondrium separate from the both the ovaries. Further investigation with a CT scan of the abdomen revealed a large well defined thin walled cystic space occupying lesion in left lumbar region extending inferiorly upto left iliac fossa and measuring 152x113mm displacing the descending colon anteriorly and to right (Fig. 1,2) . Uterus and both ovaries appeared normal.

Diagnostic laparoscopy was performed to further evaluate the mass. The mass was visualised in the retroperitoneum and procedure was converted to laparotomy. The retroperitoneal mass was dissected free from surrounding structures and removed intact. The patient made an uncomplicated post-operative recovery.

Gross pathology demonstrated a smooth well circumscribed cyst measuring 15x12 cm filled with gelatinous material. The inner surface was smooth. Microscopic examination reported mucinous epithelium of endocervical type with no evidence of atypia or invasion (Figure 3,4). The final impression of primary retroperitoneal mucinous cystadenoma was given.



Fig-1



Fig-2

Figure 1&2 : CT scan revealed large cystic lesion in left iliac fossa.

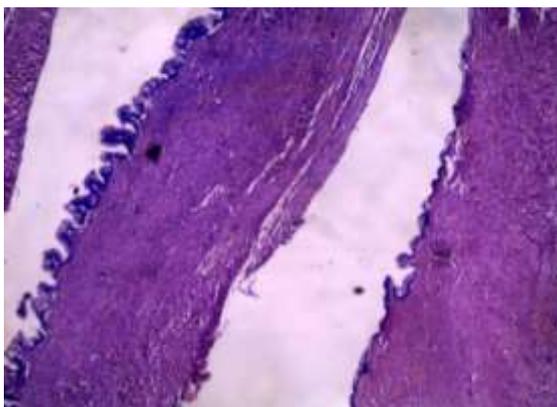


Figure 3: H & E (20X) – Section show thin wall cyst lined by columnar mucinous cells.

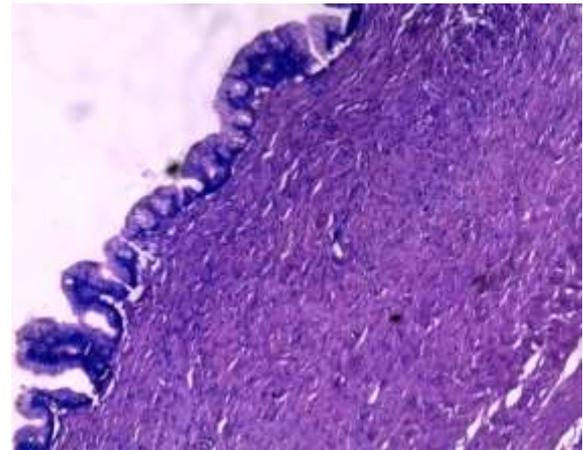


Figure 4: H& E(40X)- Section show cyst lined by single layer of columnar endocervical type mucinous cells without atypia or invasion.

DISCUSSION

Primary retroperitoneal tumour of mucinous type are extremely rare and can be further subdivide into benign, borderline or cystadenocarcinoma [3, 4]. The first description of retroperitoneal mucinous cystic tumour was undertaken by handfield jones in 1924 in his study on retroperitoneal cyst [5]. As the number of reported cases are limited, the biological behaviour and histogenesis of such tumour remain-speculative [6, 7]. Primary retroperitoneal mucinous tumour shares a histological similarity to ovarian mucinous cystadenomas but can arise at any location in the retroperitoneum without attachment to the ovary [7]. Mucinous cystadenomas are more commonly found in the pancreas and ovary. Since the retroperitoneum does not contain mucinous epithelium, therefore the origin of mucinous cystadenoma in the retroperitoneal is widely debated and remains unclear [8]. Although four main hypothesis have been given. According to the first three hypothesis, the tumour arise either from ectopic ovarian tissue, from a teratoma in which the mucinous epithelium has overridden all other components to survive as single cell component, or from remnants of the embryonic urogenital apparatus [9]. Recently a fourth theory has gained wide acceptance, the theory suggests coelomic metaplasia as the causal agent where by tumour arise from invagination of peritoneal mesothelial layer that undergoes mucinous metaplasia with cyst formation [10].

To our knowledge only 35 cases of primary retroperitoneal mucinous cystadenoma (PRMC) have been published in the English literature. All previous case were women aged 14-85 years with the mean age 39 year [11, 12]. Subramony et al. showed that histologic, immunohistochemical and electron microscopic examination of PRMC lining epithelium cells share features of mesothelial cells in addition to ovarian mucinous cystadenoma [13]. They also reported that estrogen receptor is positive in stromal cells of

PRMC which could explain the exclusively occurrence of these tumour in women [13, 14]. Although, three cases of PRMC in men have been reported in literature [14]. But some author questioned the diagnosis of these lesion as benign because all three patients died of the disease [16].

Usually this kind of retroperitoneal tumor is asymptomatic. Like most retroperitoneal mass, they cause symptoms when growing large enough to exert pressure or obstruction effect on adjacent organ [7]. However, the most common symptoms are abdominal mass, distention, discomfort or pain. Early diagnosis of primary mucinous cystadenomas is important because of their malignant potential.

Diagnosis of retroperitoneal tumours is important but difficult as serological investigations, ultrasonography, computed topography and magnetic resonance imaging, although useful, cannot allow a confident diagnosis [17]. However Motoyama *et al.* reported that measurement of CEA level in the cystic fluid may be useful in making the diagnosis [18].

Primary retroperitoneal mucinous cystadenoma manifests as a homogeneous, unilocular cystic mass at CT scan. It is difficult to differentiate this mass from cystic mesothelioma, cystic lymphangioma, and nonpancreatic pseudocyst [9]. Although aspiration is a good method to delineate the nature of the cyst, cytologic analysis of the aspirated fluid often fails to reveal the type of epithelial cells lining of the cyst. Therefore, exploratory laparotomy with complete excision of the cyst is usually indicated for both diagnosis as well as treatment [19]. Histopathological analysis of the cyst reveals a single layer of tall columnar epithelial cells with pale cytoplasm and basal nuclei. However the prevention of cystic fluid spillage during laproscopic manipulation is important especially when pathology of the retroperitoneal cyst is unclear [20]. As for management of PRMC, complete surgical excision is recommended to eliminate the risk of infection, recurrence and malignant transformation. Exploratory laprotomy with complete enucleation of the cyst is traditionally indicated however laproscopic excision of a PRMC has been reported.

CONCLUSION

Primary retroperitoneal mucinous cystadenoma is a very rare tumour. It should always be included in the list of differential diagnosis when confronted with a cystic mass in the retroperitoneum. Complete surgical removal of the tumor should be done because of the high risk of infection, recurrence and malignant potential.

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