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Case Report

Mucinous cystic neoplasm of the spleen with ovarian like stroma: a rare entity

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ABSTRACT

Mucinous cystic neoplasms (MCNs) of the spleen are very rare, with a few reported cases in the literature. They are often confused with more common benign or parasitic splenic cysts due to very similar imaging features. We report a case of a 52-year-old woman with a giant multicystic lesion in the spleen, initially thought to be a hydatid cyst. Contrast-enhanced CT revealed a giant, cystic lesion. Surgical splenectomy was performed, and histopathology confirmed a mucinous cystadenoma with an ovarian-like stroma. Immunohistochemistry revealed estrogen and progesterone receptor positivity, confirming MCN. This case demonstrates the importance of considering MCN in the differential diagnosis of large, complex splenic cysts, even without pancreatic involvement. The presence of septations, wall calcifications, mass effect and absence of daughter cysts on imaging should raise suspicion for MCN. Histopathological evaluation remains gold standard, since preoperative imaging cannot reliably distinguish these rare neoplasms. Complete surgical resection is curative and essential due to the potential risk of malignant change. Awareness of splenic MCN is important for accurate pre-operative planning and definitive surgical management.

Keywords: Mucinous cystic neoplasm, Ovarian-like stroma, Splenic cyst, Mucinous cystadenoma, Splenectomy

INTRODUCTION

Mucinous cystic neoplasms (MCNs) are well-defined in pancreas but are extremely rare in extrapancreatic sites like liver, mesentery and spleen. The 5th edition of world health organization (WHO) classification of digestive system tumors describes ovarian-type stroma as a defining histological feature of MCNs, irrespective of their location.[1] Primary splenic cystic lesions account for around less than 1% of all splenic pathologies, and are mostly congenital, post-traumatic, infectious, or neoplastic.[2]

MCNs of the spleen are especially rare, with less than 20 documented cases globally.[3-6] Distinguishing these lesions from more common pathologies like hydatid cysts or epidermoid cysts remains a major diagnostic challenge.

MCNs are well-defined cystic epithelial neoplasms that affect middle-aged women, which commonly arises in the pancreas, where they account for a particular subgroup of cystic pancreatic tumors.[1] On histological examination, MCNs are characterized by mucin-producing columnar epithelial cells supported by a subepithelial, densely cellular ovarian-type stroma, which is immunohistochemically positive for estrogen and progesterone receptors. This stroma is considered a pathognomonic feature. Due to their rarity and nonspecific signs and symptoms, splenic MCNs are often wrongly diagnosed preoperatively as mostly splenic cystic lesions like epidermoid cysts, hydatid cysts, lymphangiomas or post-traumatic pseudocysts.[7,8] Among primary splenic cysts, true epithelial cysts are a minority, and neoplastic cysts like MCNs are even uncommon.[8] The patient may present with abdominal fullness, pain, or a palpable mass when the lesion attains a significant size.

Imaging modalities such as USG, CT and MRI can identify cystic features, septations, calcifications and internal complexity, but these findings are not pathognomonic and may overlap with parasitic or benign congenital cysts.[10,11]

Being a premalignant lesion as documented in pancreatic MCNs, 10-30% may show malignant transformation hence early identification and surgical excision are important.[12]

CASE REPORT

A 52-year-old female patient presented to our OPD with a history of abdominal bloating of 2 years duration and left upper abdominal discomfort of 1 year duration. She was recently diagnosed with type 2 diabetes mellitus with an HbA1c 9.2% and also had a history of tobacco use. There was no history of trauma, fever, or weight loss. On physical examination a non-tender mass was noted in the left hypochondrium. Contrast-enhanced CT abdomen revealed a 17.5×14.6×20.6 cm multiloculated cystic lesion arising from the spleen with internal septations and wall calcifications (Figure 1). No solid components or daughter cysts were present. Pancreatic atrophy and displacement of surrounding structures were noted. Serum IgG for *Echinococcus granulosus* was negative, hence hydatid disease was ruled out. The patient underwent an open splenectomy after optimization of glycemic control. Intraoperatively a large, well encapsulated cystic mass was noted confined to the spleen (Figure 2).

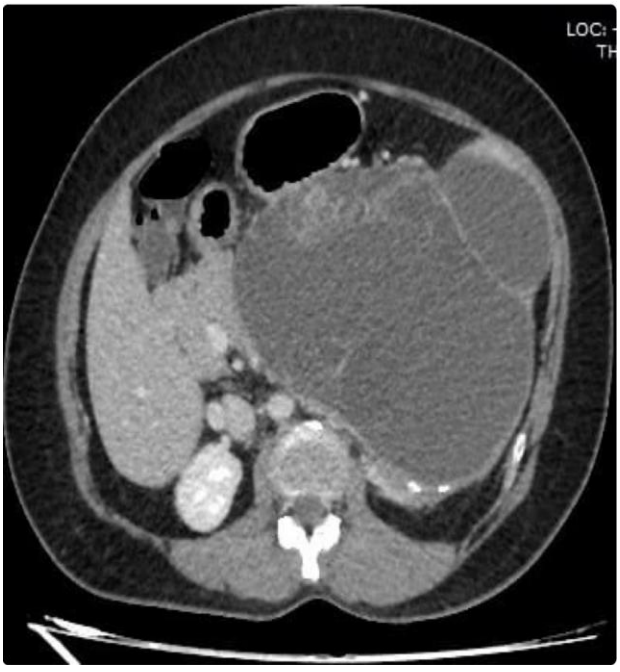

Figure 1: Contrast enhanced CT scan of abdomen showing large multicystic lesion in the spleen, displacing surrounding organs like the pancreas and aorta.

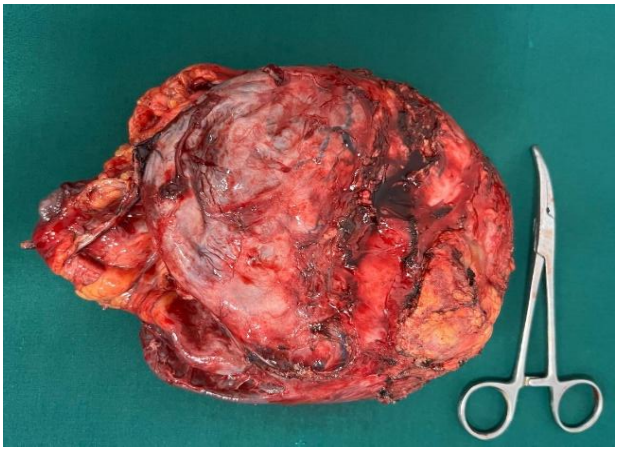

Figure 2: Gross Surgical specimen.

Resected specimen was a multiloculated cystic lesion of size 18×18×10 cm (Figure 3). Microscopically the lesion was lined by mucin-secreting columnar epithelium, with papillary projections. The underlying stroma was densely cellular which was characterized by spindle-shaped stromal cells with bland, vesicular nuclei (Figure 4). The surrounding splenic parenchyma showed chronic venous congestion with numerous Gamna-Gandy bodies, suggestive of long-standing vascular stasis. Immunohistochemical analysis demonstrated strong nuclear positivity for estrogen receptors (ER) (Figure 5) and progesterone receptors (PR) (Figure 6). These typical histopathological and immunohistochemical features were consistent with diagnosis of a mucinous cystadenoma with ovarian-like stroma. Following the enbloc resection of lesion patients symptoms subsided and recovered well. The patient was under followup for next 6 months monitoring her symptoms and no recurrence were observed.

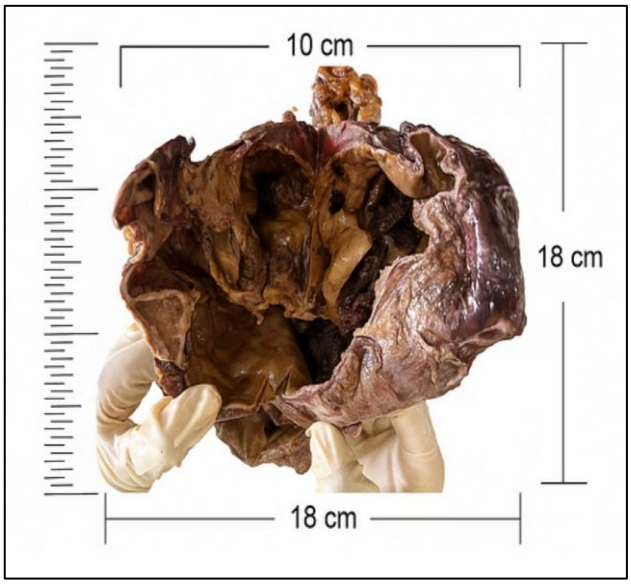

Figure 3: Resected spleen showing a large multicystic mass with multiple loculations and thick fibrotic walls.

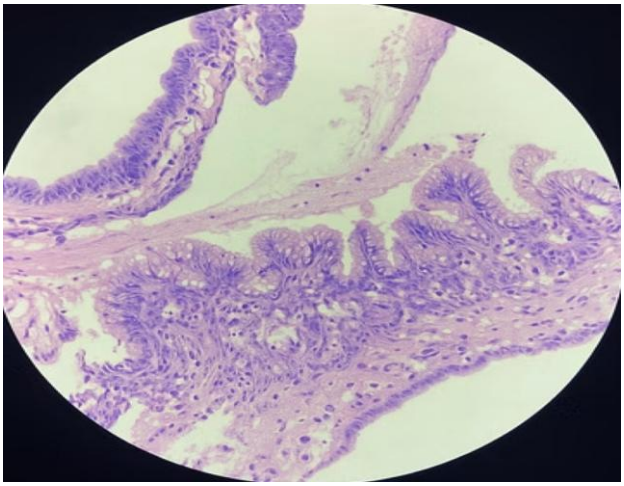

Figure 4: H and E-stained section: Histopathological image showing mucin secreting columnar epithelium and ovarian like stroma in a splenic lesion.

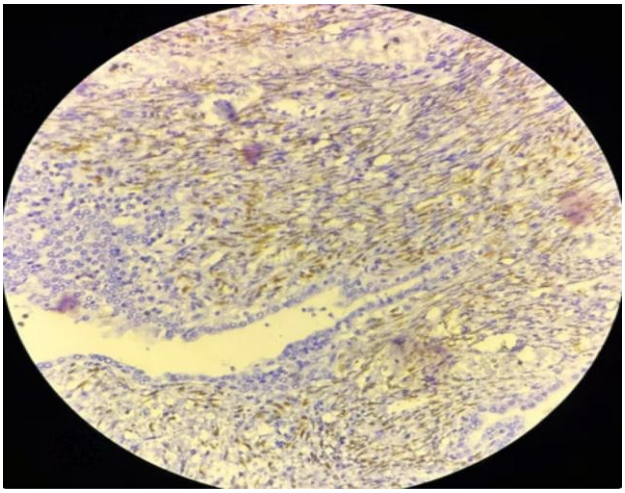

Figure 5: ER immunohistochemistry-image showing estrogen receptor positivity in splenic MCN.

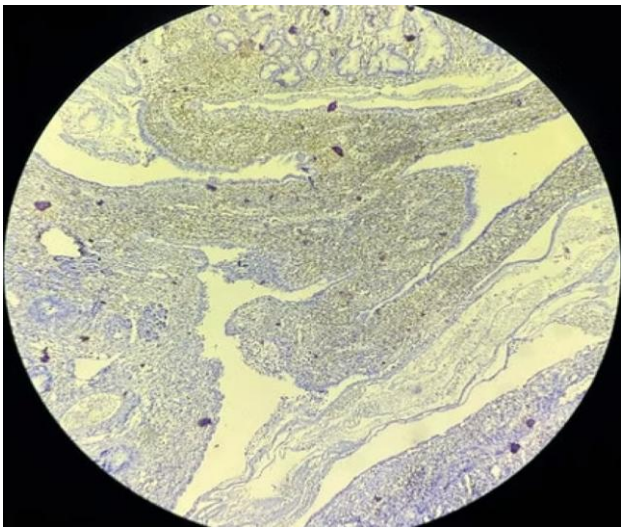

Figure 6: PR immunohistochemistry-image showing progesterone receptor positivity in splenic MCN.

DISCUSSION

MCNs are most commonly seen in the pancreas, with a high potential for malignant transformation. However, the occurrence of MCNs is extremely rare in extrapancreatic locations like spleen. Till date, only a handful of cases have been reported in the world from countries like The United States, Korea and Japan.[1,4-6] As reported by Tsuboi et al in a clinicopathologic review of over 400 splenic cysts, none of the cases were diagnosed as mucinous cystadenomas with ovarian-like stroma. It highlights the extreme rarity of this lesion in spleen.[7,8]

The diagnosis of splenic MCNs are usually delayed or missed due to their vague clinical presentation and closer differential of other cystic lesions of the spleen like parasitic and congenital cysts. These patients usually present with vague abdominal discomfort, bloating or a palpable mass, symptoms that are not pathognomonic. In our case, the patient presented with progressive abdominal distension and chronic left upper abdominal pain without any systemic symptoms or risk factors suggestive of malignancy.

The primary differential diagnoses for cystic lesions of the spleen are hydatid cysts, epidermoid cysts, and pseudocysts.[15] Hydatid cysts, caused by *Echinococcus granulosus*, are endemic in certain regions and typically present with daughter cysts on imaging (depending on the stage) and usually with a positive serology; these were not present in our case. Epidermoid cysts are usually unilocular and lined by squamous epithelium, and pseudocysts-often post-traumatic-after an episode of acute pancreatitis.[13] While contrast-enhanced CT and MRI can provide detailed characterization of cystic composition (including multiloculation, septations, and calcifications), they cannot reliably differentiate MCNs from other cystic lesions of the spleen, especially when no solid mural nodules or internal debris are noted.[6]

Thus, histopathological examination remains the gold standard for definitive diagnosis. As per the 5th edition of the world health organization (WHO) classification of digestive system tumours, the diagnosis of MCN requires the presence of a 1) mucinous epithelial lining, 2) subepithelial ovarian-type stroma, and 3) positivity for estrogen and/or progesterone receptors (ER/PR) on immunohistochemistry.[1] Our case fulfilled all of these criteria: the cyst was lined by mucin-secreting columnar epithelium with papillary projections and stained positive for ER/PR-positive ovarian-like stroma. The pathogenesis of extrapancreatic MCN are poorly understood, but the presence of ovarian-type stroma supports a theory of origin from ectopic Müllerian remnants, potentially due to embryological misplacement or metaplastic transformation of mesothelial cells.[14]

Although our case was histologically benign, it is important to recognize MCNs as they carry a risk of malignant transformation, particularly when they attain

large sizes or exhibit mural nodules or cytological atypia. In the pancreas, up to 30% of MCNs undergo malignant transformation.[14] Such transformation has not been studied with definite associations in splenic MCNs due to their rarity, the theoretical risk justifies complete surgical excision as the treatment of choice.[12] Thus, a definite preoperative plan is to be kept in mind given that preoperative imaging cannot reliably exclude the presence of in situ or invasive carcinoma within the cyst wall.

Surgical management via splenectomy is thus the recommended standard. While laparoscopic splenectomy may be considered for small, benign-appearing lesions, but larger cysts-like the 17.5×14.6×20.6 cm mass in our patient-typically require open surgical resection to avoid intraoperative rupture, spillage, and potential dissemination.

The prognosis following complete excision of benign splenic MCNs is excellent, with no reported cases of recurrence to date. However, long-term follow-up with imaging is advisable to monitor for delayed recurrence.[9] This case, one of the largest documented splenic MCNs in the literature, stresses on the importance of maintaining a high index of suspicion for rare neoplastic entities in patients presenting with large, complex cystic splenic lesions.

CONCLUSION

This case will add to the limited literature on splenic mucinous cystic neoplasms and it stresses on the importance of a high index of suspicion in evaluating complex splenic cysts. Surgical resection provides cure and avoid potential malignant transformation. Histopathology and immunohistochemistry are the final end for diagnosis.

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