

“MUCINOUS ADENOCARCINOMA OF COLON MIMICKING CYSTIC LYMPHANGIOMA: REPORT OF AN UNUSUAL PRESENTATION WITH BRIEF REVIEW OF LITERATURE.”

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ABSTRACT

Retroperitoneal masses present with a diverse group of lesions; ranging from benign to malignant. These lesions can be solid or cystic, single or multiloculated, and the contents may range from serous to mucinous fluid. Mucinous adenocarcinoma of retroperitoneum is rare. We present a case of retroperitoneal mucinous adenocarcinoma of colon in a 25-year-old male, mimicking a benign cystic lesion on radiology. Cytology along with histopathology helped in making a definitive diagnosis of malignancy in this case.

KEYWORDS : Cystic lesion, FNAC, gelatinous fluid, histopathology, lymphangioma, mucinous adenocarcinoma.

INTRODUCTION:

Retroperitoneal mucinous adenocarcinoma (RMA) is extremely rare and the histogenesis of this tumor remains unknown. In these cases, clinical symptoms are perceived by patients only when the mass grows to a sufficiently large size. Laboratory studies usually fail to detect these lesions and imaging methods merely reveal cystic lesions, neither of which result in accurate diagnosis. Surgical resection is standard for the treatment of RMA. We present a case of mucinous adenocarcinoma in a young male presenting with a large abdominal mass along with a mass in splenic flexure, which clinically and radiologically mimicked as a cystic lesion.

CASE REPORT:

A 25-year-old male presented with pain and lump in left upper abdomen for last 6 months. He also had recurrent episodes of vomiting for last 1 month. There was history of anorexia and weight loss in the patient. The patient had history of pulmonary Koch's 8 years back and abdominal Koch's 1 year back for which he had completed the antitubercular therapy. On general examination, the patient was well oriented with stable vitals. However, pallor was noted. Per abdominal examination revealed tensed abdomen with fullness present in the left side of umbilicus. A palpable lump measuring 10x10 cms was noted in left lumbar & left periumbilical region. Ultrasound abdomen revealed a well-defined heteroechoic mass lesion measuring 16x8 cms in the infraumbilical region with vascularity. The mass also extended into the left pararenal space. CECT abdomen showed a large abdominal lobulated cystic hypodense lesion in left lumbar region, abutting the descending colon, with thickening of adjacent descending colon and colo-colic intussusception and intraluminal soft tissue component [Figure 1 a-b]. Radiologically, differential diagnosis of cystic lymphangioma or any mitotic lesion was considered.

Ultrasound guided fine needle aspiration cytology was done from the left abdominal mass and yielded thick gelatinous aspirate. Smears prepared were cellular comprising of tumor cells in background of mucin. These cells revealed mild to moderate pleomorphism, and arranged in sheets, clusters, groups and papillaroid fragments, cohesive ball like clusters and singly scattered [Figure 2a-c]. These cells showed moderate anisonucleosis with focal overcrowding & overlapping and central to eccentrically placed round to oval nuclei, granular chromatin, prominent nucleoli and moderate

to abundant of pale, vacuolated cytoplasm [Figure 2d-e]. Occasional cell with marked cytoplasmic vacuolation and nucleus pushed to periphery was noted. Focal areas of necrosis and mitosis were also seen. Based on cytomorphological details, a diagnosis of mucinous adenocarcinoma was made, and biopsy correlation was advised. Colonoscopy was done and revealed a polypoidal mass at splenic flexure, obstructing the whole lumen. Cytological diagnosis was subsequently confirmed on biopsy of the abdominal mass showing multiple necrotic fragments along with focal mucin and interspersed loose cohesive clusters of atypical cells arranged in complex glandular pattern [Figure 2f]. These cells showed moderate amount of cytoplasm, hyperchromatic nuclei with prominent nucleoli. A final diagnosis of retroperitoneal mucinous adenocarcinoma (intestinal origin) was given.

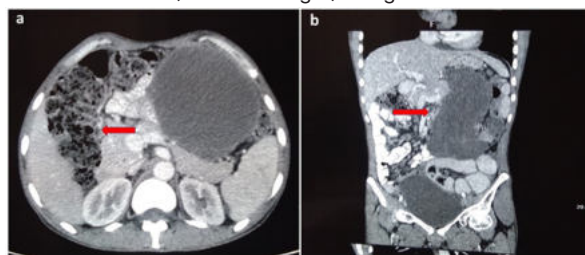


Figure 1 a-b: CECT abdomen showed a large abdominal lobulated cystic hypodense lesion in left lumbar region, abutting the descending colon, with thickening of adjacent descending colon and colo-colic intussusception and intraluminal soft tissue component.

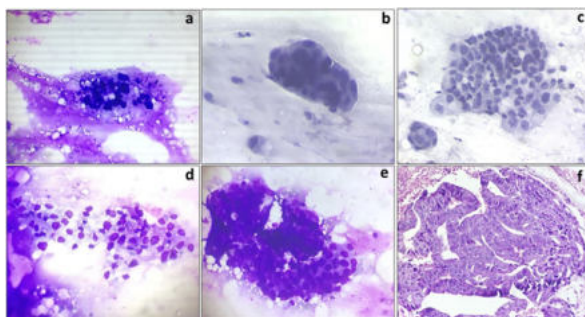


Figure 2a-f: Tumor cells noted in background of mucin; cells revealed mild to moderate pleomorphism, and arranged in

sheets, clusters, groups and papillary fragments, cohesive ball like clusters and singly scattered [α-c, α- Giemsa, 100X; b&c- Pap, 400X]. Cells showed moderate anisonucleosis with focal overcrowding & overlapping and central to eccentrically placed round to oval nuclei, prominent nucleoli and moderate to abundant of pale, vacuolated cytoplasm [2d-e, d&e-Giemsa, 400X]. Biopsy showed focal mucin, necrosis and interspersed loose cohesive clusters of atypical cells arranged in complex glandular pattern with cells showing hyperchromatic nuclei and prominent nucleoli [2f, H&E- 400].

DISCUSSION:

Retroperitoneal masses are a diverse group of lesions that range from benign to malignant. These lesions can be solid or cystic, single or multiloculated, and the contents may range from serous to mucinous fluid^{1,2}. Some nonneoplastic lesions include pancreatic pseudocyst, non-pancreatic pseudocyst, lymphocele, urinoma, and hematoma, while neoplastic lesions include cystic lymphangioma, mucinous cystadenoma, cystic teratoma, cystic mesothelioma, Mullerian cyst, epidermoid cyst, bronchogenic cyst, cystic change in solid neoplasms, pseudomyxoma retroperitonei and perianal mucinous carcinoma, as well as primary pancreatic tumors, including mucinous types^{2,4}. The clinical implications and treatment options vary depending on the diagnosis, and therefore, differentiating these lesions is imperative^{3,5}. Clinical and surgical history along with radiographic results should be reviewed and considered.

Retroperitoneal primary mucinous adenocarcinoma (RPMA) is very rare phenomenon. It is commonly seen in females. Four main hypotheses have been proposed to explain the histogenic origin of the tumor. One hypothesis suggests that the tumor arises from a teratoma with predominant mucinous epithelium, whereas other authors postulate that it is caused by intestinal duplication, also known as enterogenous genesis. The third hypothesis supports that the tumor arises from heterotopic ovarian tissue. A fourth hypothesis became widely accepted, which suggests that tumors arise from invagination of the peritoneal epithelium and undergo metaplasia during embryonic growth^{1,4}. In the present case, initially mass appeared to be primary retroperitoneal lesion on CECT abdomen and PRMA on cytological and histological evaluation. However, colonoscopy revealed an intestinal origin of this lesion.

Usually, RMA occur in middle aged individuals, however, can be seen in young patients, as in our case. According to the literature, RPMA symptoms are non-specific, with the most common ones being abdominal discomfort and palpable asymptomatic mass^{4,7}. Ultrasonography, CT and magnetic resonance imaging are often used to localize the tumor. However, these methods cannot easily differentiate between a benign and a malignant neoplasm. Yang *et al*² suggested that when encountering a cystic lesion with the characteristic of displacing the colon, kidney or ureter medially, surgeons should include RPMA in the preoperative diagnosis. Needle biopsy may also be an unreliable method with which to diagnose this tumor, since it is not effective in determining malignancy in cystic tumors. However, in the present case, biopsy supported the FNAC findings.

Laparotomy is necessary to facilitate accurate decision-making and treatment. Investigators agree regarding the complete removal of the lesion^{5,8}. However, how extensive the surgery should be remains controversial^{7,9}. Preoperative diagnosis of RPMA remains difficult and surgeons should be aware of this tumor when encountering a large retroperitoneal cystic mass^{3,9}. Treatment of RPMA remains controversial. Extirpative surgery is currently the standard treatment, since the role of chemotherapy for the treatment of RPMA has yet to be determined.

CONCLUSION:

We conclude that unusual presentation of adenocarcinoma of colon should also be considered in large abdominal cystic masses. Radiology in such cases may not be conclusive and ultrasound guided FNAC may provide diagnostic clue for further management of the patient.

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