Original Research Paper

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General Surgery

CHRONIC MYELOID LEUKEMIA IN A PATIENT WITH ABDOMINAL COCOON SYNDROME: A RARE CLINICAL PRESENTATION

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ABSTRACT Described for the first time almost a century ago, Sclerosing Encapsulating Peritonitis (SEP) is characterized by development of membrane like structure around abdominal viscera, especially small bowel. It is a recognized, rare cause of intestinal obstruction. Though several malignancies have been described as risk factors for SEP, contribution of these are on the lower side and most of these are abdominal malignancies if not all. Association of leukemias or lymphomas with SEP could not be found in literature. Our patient is an 18-year-old boy, who presented with complaints of abdominal pain and vomiting. He was managed conservatively, and relevant blood investigations and imaging was done. Ultrasound of abdomen and subsequent Contrast Enhanced CT scan of abdomen confirmed SEP, whereas blood investigations confirmed diagnosis of Chronic Myeloid Leukemia. Chronic myeloid leukemia predominantly occurs in elderly males with mean age of 65 or more. CML in young patient is a rare occurrence, that coupled with occurrence of SEP makes this case unique.

KEYWORDS :

Introduction

Intestinal obstruction is a common entity faced by surgeons around the globe. Presentation of patients with bowel obstruction can range from slight abdominal discomfort in an ambulatory patient to severely dehydrated moribund patients. Severity of the disease and its management depends to a large extent on findings on examination and performance status of patient. Nevertheless, identifying the etiology of obstruction contributes significantly to immediate management as well as follow-up in these patients. [1]

Thorough knowledge regarding the risk factors and clinical manifestation of various causative factors along with increasing availability of imaging modalities has made it easier for the treating physician. However rare causes of bowel obstruction can still pose a challenge to the best of surgeons around the globe preoperatively, else they may come as accidental encounters intra operatively. One such rare entity is Sclerosing Encapsulating Peritonitis (SEP). [2]

Described for the first time almost a century ago, SEP is characterized by development of membrane like structure around abdominal viscera, especially small bowel. Etymology of the term is based on gross and microscopic findings: sclerosing collagenous tissue formed intraperitoneally that encapsulates the small bowel associated with inflammation in newly formed membrane ~ peritonitis. These patients can present with varying degrees of intestinal obstruction ranging from acute, subacute, or chronic intermittent. Patients can be asymptomatic or maybe misdiagnosed with other causes of bowel obstruction.

SEP can be primary (idiopathic) or secondary. Secondary peritonitis has several risk factors, most common of which is prolonged peritoneal dialysis. Though several malignancies have been described as risk factors for SEP, contribution of these are on the lower side and most of these are abdominal malignancies if not all. Association of leukemias or lymphomas with SEP could not be found in literature.

Over 50 percent of SEP is diagnosed intraoperatively and can be a surprise for the operating surgeon. However, knowledge of risk factors, high index of clinical suspicion and improving imaging modalities can aid in making preop diagnosis as well as avoid unnecessary surgical exploration in these patients. [2,3]

Case Presentation

Patient is an 18-year-old boy, who presented with complaints of pain abdomen, which was generalised over past 3 weeks. It was associated with abdominal distension and obstipation since past 2 days. On examination there was tenderness all over the abdomen however there was no guarding or rigidity. Bowel sounds were not exaggerated and there was no ballooning of rectum on digital rectal examination.

X ray abdomen revealed multiple air fluid levels in supine and erect position, suggestive of small bowel obstruction. However, Ultrasound (USG) of abdomen revealed: centrally clumped up bowel loops with an echogenic sheath covering the loops, along with multiseptated gross ascites and dilated bowel loops, suggestive of encapsulating sclerosing peritonitis.

With an impression of subacute obstruction of intestine due to SEP, patient was managed expectantly in surgical ward with nasogastric tube insertion, bowel rest and IV fluids. High osmolar, water soluble radio contrast (gastrografin, 40ml) was given through Ryles tube, which has proven diagnostic and therapeutic implication. Patient passed stool and flatus 10 hours after administration of dye, and subsequent Xray abdomen also concurred the finding with dye in rectum. (Figure: 1) Nasogastric tube was subsequently removed and patient was orally allowed, in a gradual manner.

To confirm SEP, contrast enhanced CT scan of abdomen was done (Figure: 2) which revealed intraperitoneal collection with smooth, thick, and enhancing peritoneal lining with clumping of small bowel loops s/o sclerosing encapsulating peritonitis with subacute small bowel obstruction. Hepatosplenomegaly was noted along with infarct of upper pole of spleen. Multiple mesenteric and retroperitoneal lymphnode enlargement present.

Complete blood count of patient which was done as part of routine follow up revealed low hemoglobin (9.4 g/dl) and marked increase in white blood cell (WBC) count (150 thousand /ml). Subsequently Peripheral smear examination of blood was done which revealed a raise in myelocyte (30%) and metamyelocyte (28%), suggestive of Chronic Myeloid Leukemia (CML) in chronic phase. BCR ABL 2 mutation was found to be positive in the patient. Patient is currently following up in hematology department for medical management for CML.

Discussion

Known by different names over the past 2 centuries, medical community around globe had recognised SEP of some type or another. Hence credit to discovery or first description of the condition could be a subject of debate. A condition known as peritoneal encapsulation (PE) was described as early as 1868 by Cleland. (2) In 1907 peritonitis chronica fibrosa encapsulate was described by Owtschinnikow, which is the first description of SEP in medical literature.(3)

PE is now understood as a developmental malformation in which an extra layer of peritoneum is formed adjunct to existing normal one from the yolksac. The accessory layer is a thin membrane formed by mesothelium. This is hence non inflammatory by nature and congenital in origin. SEP is divided into primary and secondary. It is an acquired

thick fibro collagenous membrane. Primary SEP is idiopathic and is also known as abdominal cocoon syndrome. This term was first coined by Foo et al in 1978. SEP is further classified into 4 types based on the extent of abdominal viscera covered by the encapsulating membrane. Type I and II has partial and complete encapsulation of small intestine, respectively. Type III involves intra-abdominal contents other than small bowel, like colon, appendix, or ovaries. Type 4 involves encapsulation of entire peritoneal cavity. [2]

Diagnosing a patient with SEP requires high index of clinical suspicion and identification of predisposed patients with risk factors. Xray abdomen would help in identifying intestinal obstruction with multiple air fluid levels. There maybe central clumping of bowel loops on X ray abdomen. USG of abdomen could reveal the layer of membrane that encapsulates the bowel loops as mentioned in case presentation. However, USG is operator dependent, and findings could be subjective. Contrast enhanced CT scan of abdomen is reliable imaging modality for diagnosis of SEP. Apart from describing the membrane, it can also comment regarding bowel loops, lymphnodes, calcifications, solid organs, and mesentery. It can also aid in further management by assessing need and mode for surgical intervention if required. Diagnostic laparoscopy is less commonly done when there is a diagnostic dilemma. [2]

Conventionally SEP was thought to be disease of young girls of adolescent age group. Hence initially, theories regarding etiopathogenesis of SEP revolved around retrograde menstruation and gynecological infections. Later it was found that only 25 percent of patients affected with SEP belonged to this category. Secondary SEP was associated with various risk factors most important of which is peritoneal dialysis. The inflammation due to dialysis fluid and associated infections leads to deposition of fibrin and collagen that leads to formation of membrane. Other risk factors include consumption of beta blockers, methotrexate, abdominal tuberculosis, sarcoidosis, or malignancies.

Malignancies that are reportedly associated with SEP includes ovarian, pancreatic, renal and gastric carcinoma. Only few cases have been reported so far. And all these cases being intraabdominal malignancies, suggests a local or direct effect on peritoneum rather than systemic effect. Association of leukemia and SEP could not be found in literature. [3]

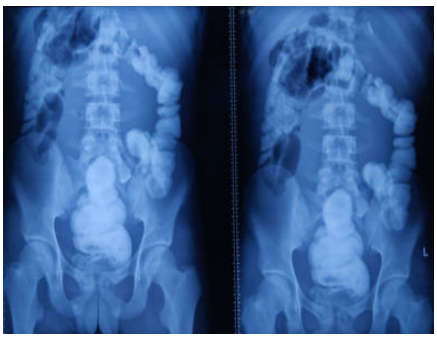
CML is a clonal proliferation of stem cells that are precursors to granulocytes. Patients are usually asymptomatic and presents with raised white blood cell count. Some patients can present with enlarged spleen, easy bruising, or malaise. Lymphoreticular organs including spleen, liver and lymphnodes could be involved, especially spleen leading to splenomegaly, infarcts and rupture.[4]

Recommendation

Round the clock availability of imaging modalities like USG and CT scan can not only aid in diagnosing conditions like SEP, but also helps in avoiding unwanted surgical explorations in these patients. Chronic myeloid leukemia predominantly occurs in elderly males with mean age of 65 or more. CML in young patient is a rare occurrence, that coupled with occurrence of SEP makes this case unique. Available data suggests that both entities are exclusive of each other. Further studies are necessary to identify any relation between the two conditions.

Figures

Figure : Xray abdomen (erect: left, supine: right) 10 hours after administering gastrograffin. Note dye in rectum (red circle) and no air fluid levels.



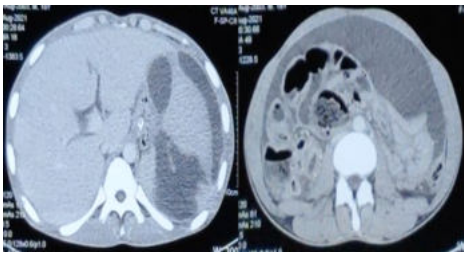


Figure : cross section imaging from CECT scan of whole abdomen, left: hepatosplenomegaly with infarct in upper pole of spleen & right: ascites with centrally clumped up dilated bowel loop suggestive of SEP with intestinal obstruction

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