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CASE STUDY

# A RARE CASE OF SMALL BOWEL PNEUMATOSIS INTESTINALIS

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**ABSTRACT**: Pneumatosisintestinalis (PI) also called as pneumatosis coli/ pneumatosiscystoidesintestinalis) is the presence of multiple gas/ air filled cysts in the bowel wall. The pathogenesis is poorly understood and is probably multifactorial. The presentation may be either an incidental diagnosis or may be associated with various gastrointestinal diseases. We report a case of 76- year- old male presented with intestinal obstruction symptoms, diagnosed to be small bowel PI associated with lymphangiectasia. In this report we emphasize the pathogenesis of PI due to lymphatic disruption.

**KEYWORDS:** Pneumatosis Cystoides Intestinalis, lymphangiectasia, Crohn's disease

## INTRODUCTION:

Pneumatosisintestinalis (PI) is a rare condition characterized by the presence of multiple gas (air) filled cysts in the bowel wall[1]. The presenting features of this condition are nausea, vomiting, abdominal pain and abdominal distension. It may be asymptomatic with an incidental radiological diagnosis. Multiple theories have been hypothesized for the pathogenesis of PI and the proven theories are bacterial, mechanical and mucosal disruption theories. PI caused by lymphatic channels disruption is not clearly understood, the etiology being increased intraluminal pressure causing leakage of gas/ air and lymph contents from the dilated lymphatics in to the bowel wall. It is otherwise known as lymphopneumatosis cystoids intestinalis or cystic lymphopneumatosis[2].

There are very few reports of PI based on lymphatic disruption pathogenesis. We present a rare case of small bowelPIassociated with lymphangiectasia .

## CASE REPORT:

A 72 year- old male presented with complaints of abdominal distension, vomiting and abdominal pain for two weeks. No history of GI bleeding, fever, jaundice or altered bowel and bladder movements were present. He underwent hemicolectomy with small bowel resection and anastomosis for terminal ileal stricture couple of months ago. Post- operatively patient had supraventricular tachycardia and was treated with beta blockers and aspirin.

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On examination: patient was conscious, oriented, and a febrile. No signs of pallor, icterus, enlarged nodes and pedal edema. Abdomen examination revealed abdominal distension, visible gastric peristalsis and right iliac fossa tenderness. Cardio vascular, respiratory and neurological examinations were normal. Renal function test showed hypokalemia. Liver function test showed marked hypoalbuminemia.

CT –enterography was done showed features of PI and air under the diaphragm. Exploratory laporatomy was done and intraoperative findings weredilated small bowel loops with adhesions. Grossly, the specimen showed multiple tiny pale brown cysts ranging in size from 0.2cm to 2.5cms over the entire length of the intestine with intervening normal serosal surface. Microscopy, showed preserved mucosal architecture and multiple cystic spaces of varying sizes in the submucosal and subserosal layers. Few spaces were lined by flattened and multinucleated giant cells. However the lining was absent in some of the spaces. The stroma was fibrous and showed scattered lymphoplasmacytic infiltrates, dilated lymphatic channels and congested blood vessels. Previous biopsy report from the stricture part of hemicolectomy specimen was small bowel lymphangiectasia with no obivious evidence of Crohn's disease (CD)such as crypt architecture distortion, submucosal granulomas and cryptitis or ischemic etiology.

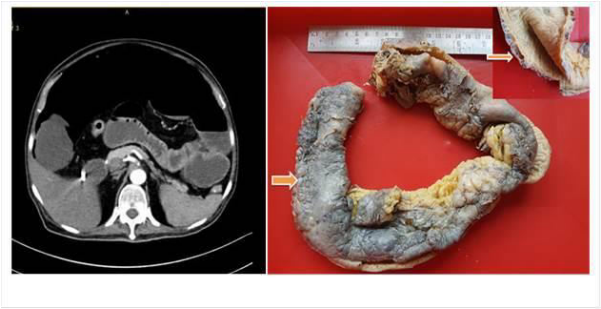

**Fig 1: CT scan showing small bowel pneumatosis and gross appearance of the small bowel pneumatosisintestinalis (PI). Inset showing sub mucosal PI, (indicated by arrow).**

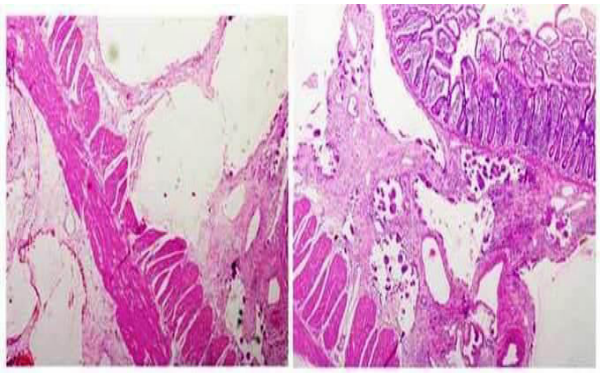

**Fig 2: Microscopic sections showing small bowel submucosal andsubserosalcystic spaces lined by multinucleated giant cells(G). (H&E x 10)**

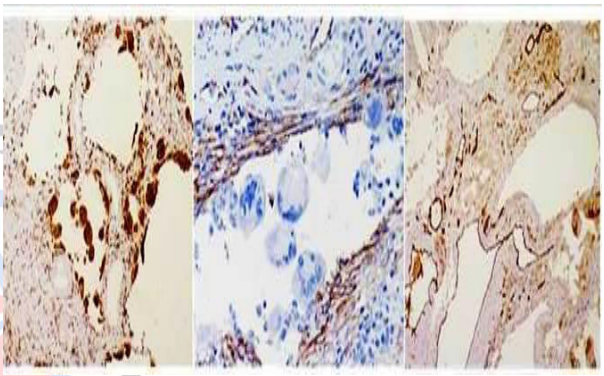

**Fig 3: Immunostain expression of CD 68, D2-40 and CD 31 on the multinucleated giant cells, Podoplanin (D2-40) immunostain expression on the pericystic interstitium and lymphatic channels respectively.**

IHC study, showeda strong linear expression (nuclear positivity) of Podoplanin (D2-40) in the pericystic interstitium and the lymphatic endothelial cells. CD68 was expressed (cytoplasmic positivity) in flattened cells and multinucleated giant cells. CD 31 immunopositivity (nuclear positivity) was observed in lymphatic and vascular endothelial cells and negative in cystic spaces.

In the immediate post-operative period patient had persistent hypotension and shock, In spite ofgood supportive care he died of cardiopulmonary arrest.

## DISCUSSION:

PI is a rare entity accounting about 0.03 to 0.37 % incidences in general population and the etiology is multifactorial. Itincludes primary/ sporadic (15%) or fulminant (85%) causes such as ischemia, infections, post-surgical or inflammatory conditions (Crohn's/ ulcerative colitis)[1].Few of these predisposing factors were noted in our case such as history of previous surgery and the coexistence of CD features.

There are very few reports of PI due to predisposing CD[5-7]. The pathogenic mechanism explained was inflammation induced mucosal injury and/or increased intraluminal pressure allowing intraluminal gas to enter the damaged bowel wall[8]. This case didn't show obvious mucosal injury favoring CD causing PI.

The other possible pathogenesis was explained by Pedicaet al[9] in their study, that there is an increased lymphangiogenesis and diffuses lymphangiectasia in CD patients when compared to their normal controls. Our patient exhibited lymphangiectasiaas the prominent finding.

Harries et al[10]stated that the congenital lymphatic abnormality directly plays a role in the development of PI. They also described about the spatial relationship of abnormal small bowel lymphatics and PI formation.

In the study done by Staudacher and Bencini[3] in the intestine of hogs clearly demonstrates the distended lymphatics forming the cystic spaces of PI. Haboubiet al[4] demonstrated the lymphatic vessel forming cystic spaces of PI by light and electron microscopy. Gui et al[2] in another study observed the expression of lymphatic marker [podoplanin(D2-40)] in the pericysticinterstitium of all cases studied, suggesting that PI is due to the rupture of the distended lymphatics .This finding is concomitant in our study thereby supporting the lymphatic theory of PI.

## CONCLUSION:

This case is unique in presentation, as the primary diagnosis itself posed a challenge. So far there has been no case reports of PI associated with lymphangiectasia. Combined clinical and radiological investigations with a high index of suspicion leads to earlier diagnosis and management of this condition otherwise a rare entity.

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