CASE REPORT

Rare presentation of benign idiopathic pneumatosis cystoides intestinalis mimicking colonic polyposis

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Introduction

Pneumatosis cystoides intestinalis (PCI) refers to the presence of air within the submucosal or the subserosal layers of the intestine. It is an uncommon condition usually secondary to other primary intestinal or extraintestinal pathologies [1].

Primary or idiopathic PCI is a rare condition that accounts for only less than 15% of all the reported cases [2]. Although Idiopathic PCI can present with or without related intestinal symptoms, the literature review showed that most of the reported cases have presented with symptoms [2–5].

Even though there are several Idiopathic PCI cases reported in world literature, we could not find a case originating from Sri Lanka. Here we are reporting a patient with Idiopathic PCI that mimicked colonic polyposis.

Case presentation

A 66-year-old male patient presented to the surgical clinic with a lump at the anus. There were no other Colorectal symptoms and he did not have any family history of colorectal cancer or polyposis syndromes. The patient did not have any co-morbidities and he was not on any long-term drugs. Proctoscopy examination revealed second degree haemorrhoids.

He underwent a screening flexible sigmoidoscopy as per the unit protocol before treatment of the haemorrhoids. Flexible sigmoidoscopy revealed multiple polypoidal lesions situated at the splenic flexure region. Subsequently, he underwent a colonoscopy which revealed multiple polypoidal lesions (>50) ranging from 5mm to 20mm (figure 1) located very close proximity to each other from mid transverse colon up to the proximal descending colon. Rest of the colon up to caecum was unremarkable.

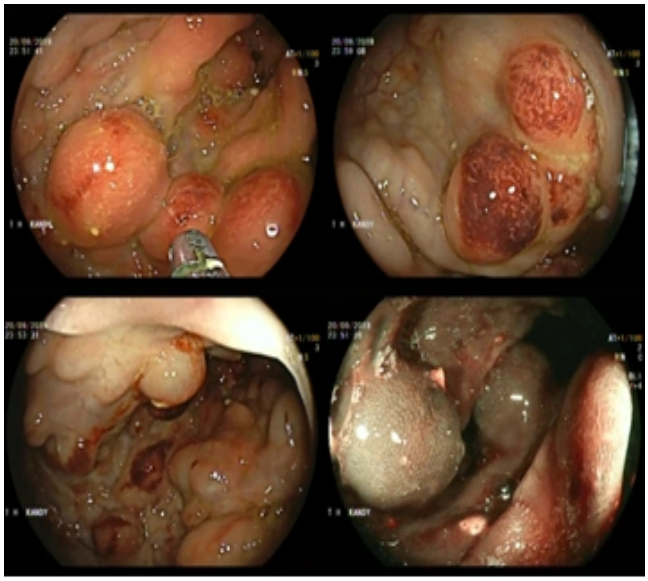

Figure 1. Multiple polypoidal lesions seen in endoscopy. Right lower image is a blue light endoscopy (BLI) image showing normal vascular pattern as the adjacent bowel

Further endoscopic examination of the polyps with Blue Light Endoscopy (BLI) was in favour of benign polyps.

Multiple biopsies were taken and one of the larger polyps were taken for histology by snare polypectomy. Histology reports from endoscopic samples revealed a benign tubular adenoma with low-grade dysplasia. Computed Tomography was not done due to a long waiting list.

As there were concerns of possible sampling error due to a large number of polyps and future risk of malignant transformation, we decided to go ahead with a left side hemicolectomy to remove and assess the affected bowel segment. The patient underwent a laparoscopic assisted left hemicolectomy. Splenic flexure, distal transverse colon and descending colon were mobilized and delivered though a mini laparotomy incision to assess the extent of the resection required. Involved bowel segment was resected completely followed by colo-colic anastomosis. The patient had a smooth post-operative recovery.

On the histological evaluation of the surgical specimen, cut sections showed air filled circumscribed polypoidal lesions ranging from 8mm to 15mm located mainly in the submucosa.

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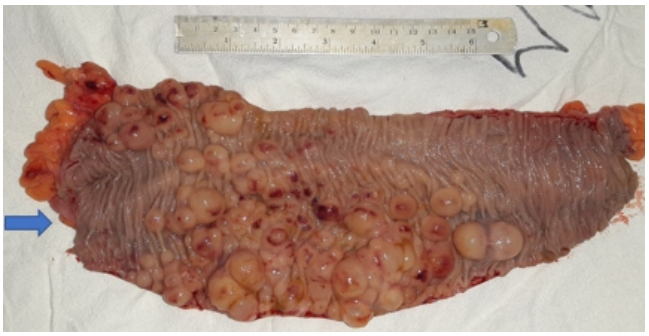

Figure 2. Surgical specimen opened longitudinally. Arrow at the distal resection margin which is well away from mucosal lesions.

Overlying mucosa showed mild architectural distortion. The lamina propria was seen to be oedematous and containing a chronic inflammatory cellular infiltrate. But there were no granulomata or any evidence of dysplasia or malignancy. The histological appearance was consistent with pneumatosis cystoides intestinalis.

Discussion

The first account of PCI appeared in French literature by Duvernoy in 1754 [6]. A number of reported cases have since then increased owing to increasing numbers of imaging investigations of the abdomen [1].

PCI should be thought of as a sign rather than a disease itself as it can be associated with life threatening causes and the patient should be carefully evaluated to identify the underlying cause. When the cause is apparent on the evaluation of the patient, it is categorized as secondary PCI which accounts for about 85% of the cases [2]. Secondary PCI can be secondary to life threatening conditions such as intestinal ischaemia, bowel obstruction, fulminant colitis, toxic megacolon, immunosuppression following organ transplantation, mesenteric vascular disease and trauma. In these situations, prompt surgical intervention is needed.

Idiopathic PCI accounts for about 10% to 15% of the cases where a cause is not apparent on further evaluation of the patient [1]. It is a benign condition which is not life threatening, thus a more conservative approach is suggested in the initial management of this condition. Even though some authors have suggested that the severity of idiopathic PCI can not be assessed based on the radiological studies, generally extensive bowel involvement, presence of portal venous gas and free peritoneal air suggests severe disease. Literature shows several instances where even pneumoperitoneum due to benign PCI have been treated successfully with conservative measures [4]. But such patients need to be carefully observed for any deterioration that would require prompt surgery.

Conservative methods of treatment include observation, hyperbaric oxygen therapy, endoscopic fine needle aspiration and antibiotic therapy. Some authors have suggested hyperbaric oxygen therapy to be the most effective means of treating benign PCI. It works by increasing the concentration of oxygen in the cyst space allowing other gases to diffuse out of the cyst and eventually collapsing the cystic space. Endoscopic fine needle aspiration aims to puncture the cyst allowing the cyst to collapse [2]. But it is associated with increased risk of infection and occasionally associated with bowel perforation as well [2].

Endoscopic diagnosis of PCI is rare as the polypoidal appearance of the lesions often misleads the endoscopist as in this case. Endoscopic ultrasound is necessary to come to an accurate endoscopic diagnosis. If the suspicion arises during the endoscopy of PCI, a CT (Computed Tomography) scan would be required to confirm the diagnosis.

We failed to come to an accurate pre-operative diagnosis mainly due to being misled by the endoscopic appearance and the initial biopsy report which suggested low grade dysplasia which was suggestive of an adenomatous lesion. Most likely the architectural changes due to inflammatory changes (as seen the final pathology report) may have led to the interpretation of initial biopsy samples as having low grade dysplasia.

Conclusion

This experience highlights the importance of having PCI as a differential diagnosis when assessing polypoidal lesions in endoscopy. It also emphasises the value to endoscopic ultrasound in evaluating such lesions. An accurate pre-operative diagnosis could have been made if the CT scan was available, thereby a less invasive treatment could have been offered to the patient.

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

References

1. Ho LM, Paulson EK, Thompson WM. Pneumatosis Intestinalis in the Adult: Benign to Life-Threatening Causes. Am J Roentgenol [Internet]. 2007 Jun 1 [cited 2020 Jan 11];188(6):1604–13. https://www.ajronline.org/doi/full/10.2214/AJR.06.1309
2. Ling F, Guo D, Zhu L. Pneumatosis cystoides intestinalis: a case report and literature review. BMC Gastroenterol [Internet]. 2019 Nov 6 [cited 2020 Jan 11];19(1):176. https://doi.org/10.1186/s12876-019-1087-9
3. Peter SDS, Abbas MA, Kelly KA. The Spectrum of Pneumatosis Intestinalis. Arch Surg [Internet]. 2003 Jan 1 [cited 2020 Jan 11];138(1):68–75. https://doi.org/10.1001/archsurg.138.1.68

4. Brill SE, Skipworth J, Stoker DL. Conservative Management of Pneumatosis Intestinalis and Massive Pneumoperitoneum in the Acute Abdomen: A Case Report. Ann R Coll Surg Engl [Internet]. 2008 Mar [cited 2020 Jan 11];90(2):W11–3. https://doi.org/10.1308/147870808X257193

5. Jacob T, Paracha M, Penna M, Arul D, Wilson J. Pneumatosis Coli Mimicking Colorectal Cancer. Case Rep Surg [Internet]. 2014 [cited 2020 Jan 11];2014. https://doi.org/10.1155/2014/428989

Learning Points:

- Importance of having pneumatosis cystoides intestinalis as a differential diagnosis when assessing polypoidal lesions in endoscopy.
- Value of multiple modalities of imaging in assessing endoscopically detected lesions when in doubt.
- Feasibility of using conservative methods of treatment including observation, hyperbaric oxygen therapy and endoscopic fine needle aspiration in treating pneumatosis cystoides Intestinalis.