A RARE CASE OF PNEUMATOSIS CYSTOIDES INTESTINALIS AFTER APPENDECTOMY

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PRESENTATION OF CASE
A female aged 42 years, presented with acute lower abdominal pain to the surgical OPD. Imaging studies showed only a fibroid uterus. Intraoperative findings showed a swollen appendix and appendicectomy procedure was performed along with hysterectomy. The specimens of resected appendix, uterus and cervix were received in the Department of Histopathology at Elbit Laboratories. Gross finding showed a yellowish solid lesion measuring 2 x 1 cms involving the tip and mid part of appendix. Microscopy showed a well-differentiated neuroendocrine neoplasm (low grade) involving tip, mid part and small part of the base. Immunohistochemistry studies using chromogranin and Ki67 markers confirmed the diagnosis. Histological findings in uterus and cervix confirmed the presence of leiomyoma.

Patient presented for routine post-operative checkup 2 months later and radiological findings postoperatively showed an ill-defined soft tissue lesion with nodular calcification seen anterior to ascending colon. A right hemicolectomy was performed. Gross findings showed a grey white ill-defined neoplasm measuring 5 x 4 x 3 cms at the ileocaecal junction. Cut sections showed few tiny slit-like spaces. Microscopy revealed many cyst spaces lined by macrophages and occasional multinucleated giant cells, predominantly in the subserosal layer. Adjacent tissue showed chronic inflammatory reparative changes (Figure 1).

Pneumatosis cystoides intestinalis is a rare disorder, characterised histologically by the presence of multiple gas filled cysts in the subserosal or submucosal wall of the large or small intestine.¹ Pneumatosis cystoides intestinalis occurring in infants is usually seen as a component of necrotising enterocolitis and often has a fatal outcome. Exceptionally, it occurs in association with multiple sclerosis. In adults, it may present as an idiopathic finding or in association with mechanical intestinal obstruction, chronic lung disease or scleroderma. When not associated with other abnormalities, it follows a chronic and indolent course and may produce signs of intestinal obstruction and lead to an incorrect radiographic diagnosis of carcinoma.² The case described here is exceptional, as it developed after surgical removal of the appendix for which an incidental carcinoid was found histologically involving the base.

PATHOLOGICAL DISCUSSION
After the first description of pneumatosis cystoides intestinalis in 1730 by Du Vernoy in autopsy specimens, these entities were named in 1825 by Mayer. Pneumatosis cystoides intestinalis diagnosis was first established by Hahn in 1899 and later its associated radiological findings were first described by Eaumann Schender in 1939.³⁴ Pneumatosis cystoides intestinalis was previously reported as occurring most commonly in the small intestine; however, recent studies have shown that the colon is affected more frequently (46 - 62%) versus the small intestine (15 - 26%) respectively with only 7% of cases affecting both.⁵⁶ Symptoms most commonly include abdominal pain, constipation and bloating as well as diarrhoea, mucous discharge and rectal bleeding.⁷⁸ Further, Wu et al found that extramural gas was most commonly localised to the submucosal layer (69.9%) as opposed to the subserosal layer with 4% of cases involving gas in both the layers.³

Pneumatosis cystoides intestinalis could be primary in 15% of cases without obvious cause or secondary to other pathology in 85% of cases like mesenteric vascular disease, necroting enterocolitis, inflammatory bowel disease and connective tissue disorder as scleroderma. Another cause is drug therapy with immunosuppressive and chemotherapeutic drugs. Also it can result as a complication of sigmoidoscopy, colonoscopy and post-surgical anastomosis.⁹ As is evident in the described case, where the development of pneumatosis cystoides intestinalis is thought to be secondary to appendicectomy.

Imaging studies which are useful in diagnosing pneumatosis cystoides intestinalis are plain abdominal x-rays, opaque enema, computerised tomography, ultrasonography, MRI and colonoscopy. Among these abdominal x-rays is the most reliable examination.¹⁰ Though radiological examination detects most of these cases, pathological examination is a must in few cases, where endoscopy biopsy is warranted. Though pathological examination is a must, only few studies however have described the microscopic features. A larger study on microscopy will probably reveal features of prognostic importance.¹¹
the second reported case in literature of pneumatosis cystoides intestinalis associated with inflammatory changes following appendicectomy after the first published case report of Chris Vendryes et al.\(^\text{13}\) The incidental finding of carcinoid tumour in the appendix also makes this case interesting and necessitates the need for surgical intervention at this point.

**REFERENCES**