

MYOEPITHELIAL HAMARTOMA OF SMALL INTESTINESistla Radha¹, Tameem Afroz²¹Consultant and HOD, Department of Anatomical Pathology and Cytology, Global Hospitals, L. B. Nagar, Hyderabad.²Senior Consultant, Department of Anatomical Pathology and Cytology, Global Hospitals, L. B. Nagar, Hyderabad.**ABSTRACT**

Myoepithelial hamartoma or adenomyoma is a rare benign tumour like lesion of the small intestine consisting of glandular structures surrounded by smooth muscle bundles. It is usually asymptomatic in adults. Majority of these lesions are located in the stomach and their location in small intestine beyond duodenum is extremely rare. First description of this lesion was given by Magnus Alsleben in 1903. Synonyms used to describe this entity are myoepithelial hamartoma and foregut choristoma. We report two cases of adenomyomas of ileum in a 66-year-old male and a 40-year-old lady in the ileum. Ileal and jejunal adenomyomas are rare and only 26 cases have been described in literature.

KEYWORDS

Hamartoma, Ileum, Myoepithelial.

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INTRODUCTION: Adenomyomatous hamartomas are very rare benign tumour like lesions of the gastrointestinal tract and the histological diagnosis is debated. These lesions are described in the stomach, small intestine and biliary tract. Other names used to describe this entity are myoepithelial hamartoma and foregut choristomas. They are either considered epithelial hamartomas or ectopic pancreatic tissue. Histologically they are composed of glandular structures surrounded by smooth muscle bundles. There are very few cases in Indian literature describing this entity. Only 26 cases are described in literature. It is important for clinicians and pathologists to be familiar with this entity to avoid inaccurate diagnosis and unnecessary surgery.

CASE 1: A 66-year-old man presented with right lower abdominal pain and vomiting. He was a diabetic and appendectomy was done 20 years ago. Abdominal examination revealed mild distension with diffuse tenderness. No mass was palpable. There was no organomegaly. Abdominal ultrasound and CT scan revealed dilated, thickened ileum. CT scan showed mural thickening of the proximal ileum. Random blood sugar was 202 mg/dL and other biochemical and haematological parameters were within normal limits. Exploratory laparotomy was performed with a diagnosis of intestinal obstruction. Per operatively terminal ileum was thickened with multiple adhesions. A hard nodule of 1.5 cm was found 70 cm from ileocolic junction. A stricture was seen in the ileum 60 cm proximal to ileocolic junction. Ileocaecal resection was done and intestinal continuity was established. He was apparently doing well in the immediate postoperative period, but developed myocardial infarction on the 3rd day and could not be revived.

CASE 2: A 40-year-old lady presented with history of a mass in abdomen and distension of 3 days' duration. There was no history of vomiting or diarrhoea. There were no constitutional symptoms. She is not a known diabetic or hypertensive patient. She gave a history of undergoing hysterectomy and right oophorectomy for dysfunctional uterine bleeding 5 yrs. back. Her routine haematological and biochemical parameters were within normal limits. Present CT scan revealed a left ovarian cyst. On examination, abdomen was soft, nontender. A mass was felt in the abdomen. It was mobile in all directions and soft in consistency. With the diagnosis of ovarian mass, she was taken up for surgery. Per operatively, a 50 x 45 cm mass was seen arising from the left ovary. On further examination, there was a 2 x 2 cm submucosal nodule in the ileum 100 cm from ileocecal junction. An ovarian cystectomy was done and a segment of ileum with submucosal nodule was also removed. She had an uneventful recovery.

Gross Examination: Revealed an ileocaecal resected specimen measuring 83 cm. Ileum measured 75 cm. Caecum measured 8 cm. 60 cm from the ileocaecal junction, there was an ulcerated stricture measuring 1.5 cm; and 70 cm from the ileocaecal junction, there was a hard nodule measuring 1.5 x 1.5 cm. Cut surface of the nodule was grey white with a trabeculated appearance. Rest of the mucosa appeared normal.

In case 2, a 2 x 1.2 cm segment of intestine was removed. Serosa was nil remarkable. On opening, there was a submucosal nodule of 1 x 0.9 cm. Cut section of the nodule was grey white.

Microscopic Examination: H&E sections through the nodules in both the cases revealed normal mucosa, submucosa revealed a lesion composed of admixture of glands and smooth muscle bundles (Fig.1A and 1B). Glands are of various sizes, some of them are cystically dilated (Fig. 1C). Lining is composed of single layer of tall columnar cells. Glandular structures were surrounded by variable number of

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smooth muscle bundles. Glandular epithelium expressed cytokeratin 7 positivity and smooth muscle bundles were strongly smooth muscle actin positive (Fig. 1D). Proliferative activity with Ki-67 was low. Both the lesions were diagnosed as myoepithelial hamartoma of ileum. Immune profile findings were also consistent with diagnosis. In case one, rest of the intestine revealed ischaemic changes. In case 2, ovarian cyst was a serous cystadenofibroma, benign.

DISCUSSION: Hamartomas are benign conditions in which abnormal location of normal tissues is seen. There are three types of hamartomas described in the ileal part of small intestine: 1. Neuromuscular and vascular hamartoma (NMVH). 2. Neuromesenchymal hamartoma (NMH). 3. Myoepithelial hamartoma also called as adenomyoma. Adenomyoma of gastrointestinal tract are rare tumour like lesions. They are commonly seen in stomach, duodenum and biliary tract. Adenomyomas of small intestine distal to duodenum are very rare, most cases of ileal adenomyomas have been single case reports. Only 26 cases have been reported in literature.¹ They are composed of glandular structures surrounded by smooth muscle bundles.² The first description of adenomyoma was given by Magnus Alsleben in 1903 in a lesion located in the stomach.³ Benign tumours of the small intestine are estimated to be 10% of all nonmalignant tumours of the gastrointestinal tract and 30% of all neoplasms of the small intestine.⁴

Adenomyomas account for 6.8% of benign tumours of small intestine. They can appear at any age but usually occur in the fifth decade with a peak incidence in the seventh to eighth decade. They may be a cause of intestinal haemorrhage or cause of intussusception.⁵ In adults, they are commonly asymptomatic, most of these lesions are incidentally discovered during surgery or at autopsy. Widely accepted pathogenesis is they are considered as either an epithelial hamartoma or type 3 pancreatic heterotopia.⁶ Adenomyomas must be distinguished from enteritis cystica profunda, pneumatosis cystoides intestinalis, hamartomatous polyp of Peutz-Jeghers syndrome, duplication cysts and adenocarcinoma.⁷ Immunophenotypically glandular epithelial cells are cytokeratin 7 positive, similar to the normal epithelial cells of pancreatic, and biliary system. Normally Ck-7 is expressed in pancreatic duct epithelium and is absent in gastrointestinal tract. Ck-20 is positive in gastrointestinal epithelium and is absent in pancreatic duct epithelium. This immunohistochemical pattern of Ck7 (+) and Ck20 (-) supports the heterotopic pancreatic theory. Epithelial cells exhibit low proliferative activity.⁸

Adenomyomas can have rare histological findings like presence of goblet cells, Paneth cells.⁹ Handra-Luca et al reported exocrine pancreas and endocrine pancreas.⁽⁸⁾

CONCLUSIONS: Adenomyomas or myoepithelial hamartomas are benign tumour like lesions which rarely occur in the small intestine. The origin of which is considered to be an epithelial hamartoma or ectopic pancreatic tissue. Majority of them are asymptomatic and found incidentally.

Symptomatic lesions are treated by segmental resection. When they occur in the valerian system it is clinically difficult to differentiate from carcinoma and may lead to needless surgeries. It is important to keep this entity in differential diagnosis by clinicians and pathologists to make accurate diagnosis and to avoid radical surgeries.

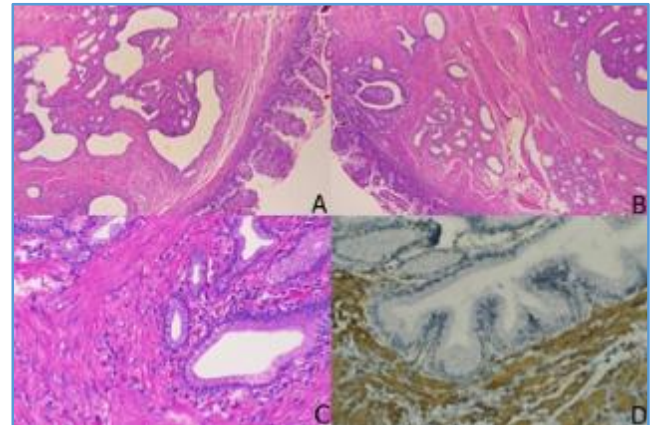


Figure 1

Fig. 1: A & B: Ileal mucosa with submucosal lesion showing admixture of benign glandular elements and smooth muscle. (H & EX100) **C:** Glands lined by single layer of columnar epithelium, separated by fascicles of smooth muscle cells. (H & EX200). **D:** Immunohistochemistry with smooth muscle actin (SMA) clearly delineating the glandular and stromal elements (x400).

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