CASE REPORT

DOI: 10.15713/ins.bhj.200

A Curious Case of Ileocaecal Mass

ABSTRACT

Background: Neuroendocrine tumours (NETs) of the ileocaecal junction are uncommon neoplasms and are frequently diagnosed incidentally. Most gastrointestinal NETs are detected on histopathological examination following surgery for presumed benign conditions such as appendicitis. Surgical resection remains the cornerstone of management, with the extent of surgery determined by tumour size, location, and pathological features.

Case Presentation: We report the case of a 46-year-old male who presented with recurrent abdominal pain, vomiting, and significant weight loss. Radiological evaluation revealed an ileocaecal mass causing luminal narrowing and features of subacute intestinal obstruction. A CT-guided biopsy suggested a low-grade neuroendocrine tumour. The patient underwent a radical right hemicolectomy. Histopathological and immunohistochemical evaluation confirmed a well-differentiated Grade 1 neuroendocrine tumour involving the ileocaecal junction, terminal ileum, and appendix, with regional lymph node metastasis.

Conclusion: This case highlights the diagnostic challenge posed by ileocaecal neuroendocrine tumours due to their nonspecific presentation. Routine histopathological examination of appendectomy and right hemicolectomy specimens is essential, as clinically occult malignancies and NETs may otherwise be missed.

Key words: Ileocaecal tumour; Neuroendocrine tumour; Appendiceal neoplasm; Right hemicolectomy

BACKGROUND

Neuroendocrine tumours (NETs) are rare neoplasms, accounting for less than 1% of all malignancies, with approximately two-thirds arising from the gastrointestinal tract.[1] Appendiceal neuroendocrine tumours were first described in 1914 by Gosset and Masson.[2] Most NETs are located at the tip of the appendix and remain clinically silent.

Definitive diagnosis relies on histopathological examination, which enables assessment of mitotic activity, Ki-67 proliferation index, and tumour differentiation, forming the basis of grading according to the World Health Organization classification.[3] We present a rare case of a primary neuroendocrine tumour originating at the ileocaecal junction.

CASE PRESENTATION

A 46-year-old male with no significant past medical history presented with generalized abdominal pain associated with nausea and vomiting. He reported multiple similar episodes over the preceding six months, accompanied by progressive weight loss and deterioration in general health.

On admission, the patient was conscious and hemodynamically stable but tachycardic, with abdominal distension. There was no fever. Laboratory investigations revealed anemia (Hb 8.3 g/dL) with a normal total leukocyte count.

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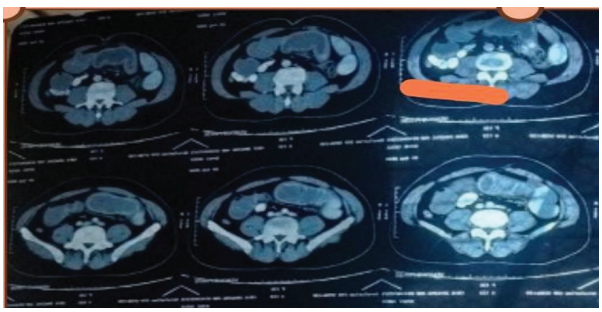
Ultrasonography demonstrated bowel wall thickening in the right iliac fossa. Contrast-enhanced CT of the abdomen revealed a moderately enhancing nodular soft-tissue lesion involving the terminal ileum and ileocaecal junction, causing significant luminal narrowing.The appendix was not separately visualized and appeared to be involved by the lesion. Associated calcified mesenteric lymph nodes were noted, raising suspicion of a neuroendocrine (carcinoid) tumour.



CT image of the ileocaecal mass.

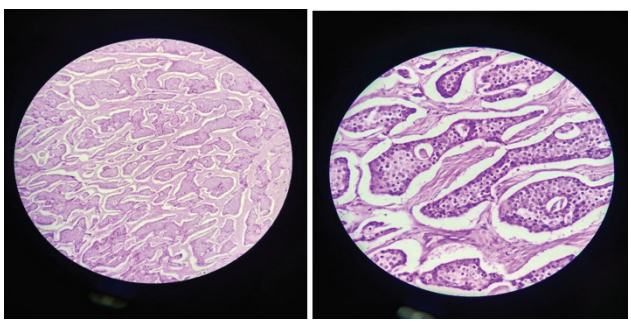
CT-guided biopsy of the lesion revealed features suggestive of a Grade 1 neuroendocrine tumour. Immunohistochemistry showed positivity for synaptophysin, chromogranin, and pan-cytokeratin, with a Ki-67 index of less than 1%.

Based on these findings, a diagnosis of ileocaecal mass causing subacute intestinal obstruction was made. Exploratory laparotomy revealed a nodular tumour involving the terminal ileum and ileocaecal junction with marked luminal compromise. A right hemicolectomy was performed. The postoperative period was uneventful, and the patient was discharged on postoperative day six.

Histopathology and Immunohistochemistry:

- Microscopy and Diagnosis:
- Tumour: Well-differentiated neuroendocrine tumour (Grade 1)
- Location: Ileocaecal junction with extension into terminal ileum and appendix
- Tumour size: 4.5 cm (maximum dimension)
- Extent: Invasion of serosa and appendiceal wall
- Mitotic rate: <2 mitoses/2 mm²
- Lymphovascular invasion: Present
- Perineural invasion: Present
- Margins: Proximal, distal, and radial margins free of tumour
- Regional lymph nodes: Metastatic neuroendocrine tumour with extranodal extension (3/6 nodes)

Pathological Stage (AJCC 8th Edition): pT4 pN1 pMx



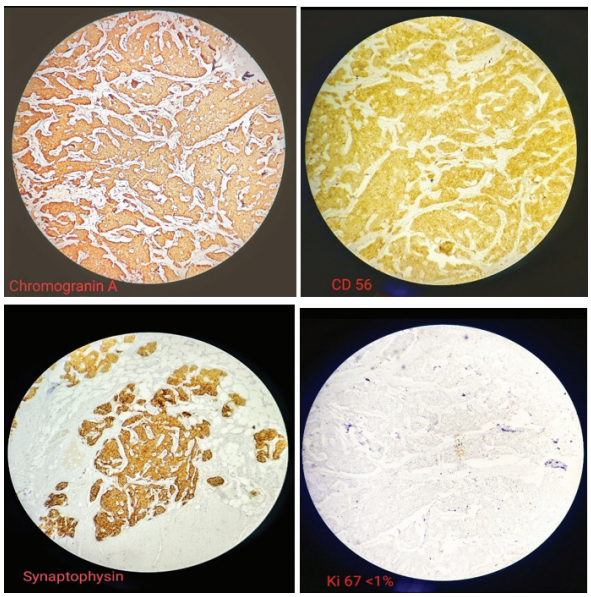
Immunohistochemistry:

- Synaptophysin – Positive
- Chromogranin – Positive
- CD56 – Positive
- Ki-67 – <1%

The immunoprofile confirmed a Grade 1 neuroendocrine tumour

DISCUSSION

Neuroendocrine tumours arise from the disseminated neuroendocrine system and share common morphological and immunohistochemical features.



Approximately 73% occur in the gastroenteropancreatic system, most commonly in the small intestine, rectum, appendix, colon, and stomach.[4,5]

Appendiceal neuroendocrine tumours have an annual incidence of 0.15–0.6 per 100,000 population, with a slight female predominance.[6] Due to their indolent nature and nonspecific symptoms, preoperative diagnosis is often difficult, and most cases are detected incidentally on histopathological examination of resected specimens.

Tumour size, location, depth of invasion, lymphovascular involvement, and proliferative index are key determinants of prognosis and surgical management. Tumours smaller than 1 cm generally require appendectomy alone, while those larger than 2 cm, involving the base of the appendix, or demonstrating aggressive features warrant right hemicolectomy with lymphadenectomy.[10,11]

Somatostatin analogues remain the mainstay of treatment for metastatic or functional tumours, while locoregional therapies may be considered for unresectable hepatic metastases.[12]

CONCLUSION

Neuroendocrine tumours of the ileocaecal region are rare and often clinically occult. This case underscores the importance of maintaining a high index of suspicion and routinely submitting appendectomy and colorectal specimens for histopathological evaluation to avoid missing potentially aggressive malignancies.

How to cite this article: Gajghate N, Shaikh MS, Mehta K. A Curious Case of Ileocaecal Mass. Bombay Hosp J 2025; 67(3):9-11.

Conflicts of Interest: None. **Source of Support:** None.

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