Ileal NET with Hepatic Metastasis

Presented By:
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Case presentation: 70 yr. old female with past medical history for Hypertension, Hyperlipedemia presents with nausea, vomiting and abdominal distension. No wbc count and normal labs.

Past Surgical History: None

Initial ED work up: CT Abdomen pelvis with contrast and MRI
CT Exam

Fig 1 a,b: Coronal and Axial post contrast CT demonstrate small bowel obstruction with a transition point in the right lower quadrant (collapsed distal ileum) which is thickened and enhancing (arrow) segment of the terminal ileum.

MRI Exam

Fig 2 c,d,e: Coronal T2 weighted MRI demonstrates small bowel obstruction and stricture of the terminal ileum (orange arrows). Images d & e axial T2 weighted and DWI B 500 respectively demonstrated T2 bright metastasis in the liver (green arrows).
• Gross Surgical Pathology image demonstrates a transition point in the terminal ileum.

• Notice the focal nodular thickening and stricture of the ileum (orange arrow).

• Upstream dilation of the obstructed bowel (green arrow).
• Cells with round to oval nuclei with salt and pepper chromatin and moderate eosinophilic granular cytoplasm arranged in nests.

Image 2: Ki-67 immunostain, 400X

• 67 labeling index is approximately 25%.

These tumor cells were also positive for synaptophysin and chromogranin-A immunostains. Chromogranin-A and synaptophysin immunostain positivity suggest neuroendocrine differentiation.

Grading system based on Ki-67 labeling index
◦ Low grade or grade 1 (G1): Ki67 index < 3%
◦ Intermediate grade or grade 2 (G2): Ki67 index 3% - 20%
◦ High grade or grade 3 (G3): Ki67 index > 20%
Gastroenteropancreatic neuroendocrine tumors (GEP-NETs)

- Distal part of the ileum is the most frequent (up to 30%) site of GEP-NETs.
- Localization of ileal carcinoids can be particularly challenging; furthermore, they are multifocal in 26%–40% of cases.

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<th>Location</th>
<th>Percentage of NETs</th>
<th>Key clinical Characteristics</th>
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| Stomach (Gastric Type 1, II and III) | Type 1: 70/80% of stomach NET  
  Type 2: 5/6% NET  
  Type 3: 15-20% NET | • Type 1: Multifocal and associated with chronic gastritis.  
  • Type 2: Multifocal and associated with ZES.  
  • Type 3: Usually solitary. |
| Small Bowel Jejunum | 22% of small bowel NET’s                    | • Early regional metastasis. Majority are well differentiated.    |
| Ileum              | 70% of Small bowel NET’s                    | • 20% with hepatic metastasis.  
  • Present with bowel obstruction and Carcinoid syndrome. |
# Imaging Modalities & Key Teaching points

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<th>Modality</th>
<th>Imaging Findings</th>
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| Multidetector CT: Unenhanced, Arterial and Venous phase. | • Hyper vascular nodular lesion or bowel thickening.  
• Desmoplastic reaction of mesentery.  
• Mesenteric nodal metastasis.  
• Enterography can demonstrate intra luminal lesions. |
| CT Enterography                |                                                                                 |
| MR Imaging                     | • MR Enteroclysis has high sensitivity to detect luminal lesions.  
• Lesions in the wall are T2 hyper intense and show enhancement.  
• DWI is sensitive to detect liver metastasis. |
| Endoscopic US                  | • Mini probes to evaluate terminal ileum.                                     |
Metastasis and Key Management steps for GEP-NETs

- Likelihood of metastases beyond the mesentery is proportional to tumor size:
  Liver Disease Risk: 20-30% in tumors <1 cm,
  over 40% in tumors >2cm
- Complete surgical resection improves 5-year survival rate with an 83% favorability and is the first line.
- Partial resection or liver transplantation is suggested to select patients as an alternative (especially in non multifocal disease).
- Local-regional cytoreductive therapies such as radiofrequency ablation embolization techniques for liver dominant symptomatic disease.
- SSTRs for well differentiated and low proliferation index tumors or metastasis.
- Systemic Chemotherapy for aggressive biology produces a response rate of 42-67%.
References:

- Ganeshan D, Bhosale P, Yang T, Kundra V. Imaging features of carcinoid tumors of the gastrointestinal tract. AJR 2013; 201:773–786
- Hicks RJ. Use of molecular targeted agents for the diagnosis, staging and therapy of neuroendocrine malignancy. Cancer Imaging 2010; 10:S83–S91