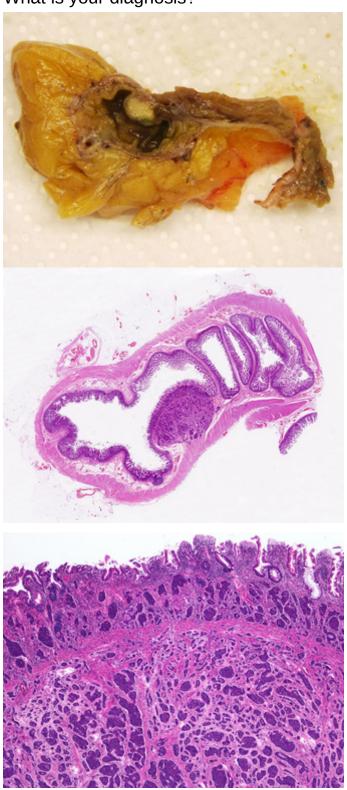
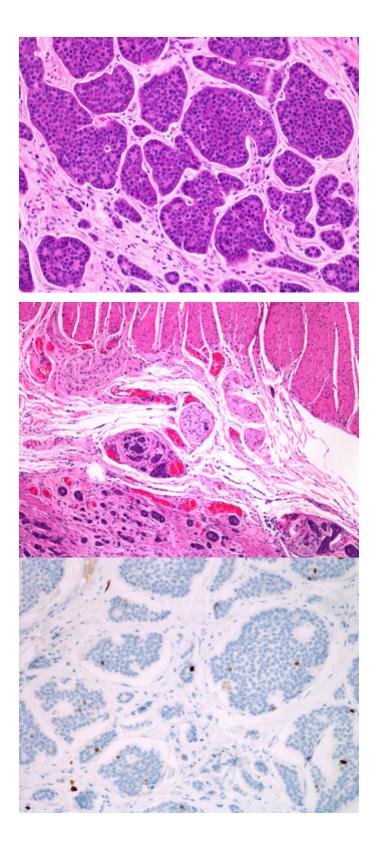
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A 56-year-old male with a tumor in the ileum and liver metastases. Resection of a small bowel segment and two liver metastases was performed. During surgery, Meckel's diverticulum was discovered and removed as well.

What is your diagnosis?





Diagnosis:

Neuroendocrine tumor (NET G1) in Meckel's diverticulum.

Comment:

In resected specimen, two similar tumors were found, one in the ileum, measuring 9 mm in diameter and another one in Meckel's diverticulum, measuring 6 mm in diameter (Panel A), both infiltrating subserosal fatty tissue. They were composed of uniform cells with few mitoses (Panels B-C) and were immunohistochemically positive for chromogranin and synaptophysin. Ki67 stained less than 2% of cells (Panel F). Perineural invasion was present in both (Panel E), whereas lymphovascular invasion was found in ileal tumor only. Metastases were found in 4/10 lymph nodes and in the liver.

Meckel's diverticulum (MD) is the most common anomaly of the gastrointestinal tract, resulting from incomplete obliteration of the omphalomesenteric duct. It is usually lined by small intestinal mucosa and may contain heterotopic gastric epithelium or pancreatic tissue. Complications include inflammation, intussusception, obstruction and bleeding. Tumors may rarely occur in MD, being present in approximately 3% of symptomatic MDs. Various tumors have been described in MD, such as NET, leiomyoma, GIST, adenocarcinoma, lymphoma, metastases, etc.

The present case is a typical case of a small bowel NET, which is well known for being able to metastasize even if very small. It also shows that NETs can be multiple, and that NET may arise in MD. In our patient it seems more likely that metastases originated from ileal tumor in which lymphovascular invasion was present. However, studies have shown that NET in MD can behave aggressively and that a significant proportion of NETs in MD had metastases at diagnosis, despite a small size and low grade of the primary tumor. It has been therefore suggested that NETs in MD must be treated aggressively as other small bowel NETs, including regional lymphadenectomy.

For further reading:

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- Lorenzen AW, O'Dorisio TM, Howe JR. Neuroendocrine tumors arising in Meckel's diverticula: frequency of advanced disease warrants aggressive management. J Gastrointest Surg. 2013; 17:1084-1091.
- van Malderen K, Vijayvargiya P, Camilleri M, Larson DW, Cima R. Malignancy and Meckel's diverticulum: A systematic literature review and 14-year experience at a tertiary referral center. United European Gastroenterol J. 2018; 6:739-747.

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