Diagnosis of Rare Duodenal Neuroendocrine Tumor with Carcinoid Syndrome

Aurika Balamurugan, Fathma Rubin Sha, Subair Sabbar Ahmed
Tbilisi State Medical University, Tbilisi, Georgia

Introduction

Duodenal carcinoids are malignant tumors originating from enterochromaffin cells. They develop distant metastases and account for approximately one-third of gastrointestinal neuroendocrine tumors (NETs). They are the primary cause of carcinoid syndrome, a prominent neuroendocrine neoplasm presentation. Primary duodenal carcinoids, accounting for less than 2% of all GI-NETs, are linked to loss of function mutations of the MEN1 gene, and the related NETs show loss of heterozygosity on chromosome 11q13.

Background

Duodenal carcinoids are rare tumors, and the secondary manifestation of carcinoid syndrome makes them a unique occurrence. The incidence is estimated to be approximately 1-2.4 cases per 100,000 population.

Case

A 34-year-old adult male presented with upper abdominal pain, diarrhea, cutaneous flushing, dry cough, exertional dyspnea, and right-sided heart disease due to regurgitant tricuspid valve deposits. Multiple nodules, both <10 mm and ≥10 mm in size, were found in the upper parts of the duodenum, along with long segment wall thickening on contrast-enhanced CT. Duodenal carcinoid elements were confirmed via positive chromogranin A in endoscopy-guided biopsy material and elevated 24-hour urine 5-hydroxyindoleacetic acid (5-HIAA). A 2-dimensional echocardiographic evaluation suggested the diagnosis of carcinoid heart disease.

Discussion

To support clinical decision-making, recent scientific publications from the last 10 years were reviewed using the keyword "duodenal carcinoid" on PubMed. One systematic review and 20 literature reviews were found. Advanced imaging tools for assessing tumor distribution, lesion size, depth of invasion, metastatic conditions, and multifocal disease in the gastrointestinal tract were discussed. The Ki67 proliferative index and depth of invasion were considered in deciding between surgical and endoscopic resection. Approaches to non-resectable metastasized NETs included Peptide Receptor Radionuclide Therapy or 90Y radioembolization as alternative treatments.

Conclusion

The patient, symptomatic with multiple tumors in the duodenum, required radical surgery and regional lymphadenectomy, as tumor size, depth of invasion, and multifocality could not reliably predict lymph nodal metastasis. The benefits of chemotherapy were not proven. The patient was subsequently referred to specialized oncology care. This case report aims to highlight the often-overlooked clinical manifestations of duodenal carcinoids to aid in early recognition and diagnosis, focusing also on the diagnostic methods employed. Large-scale research is required to better understand various aspects of this disease.