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Goblet cell carcinoids of the appendix: Tumor biology, mutations and management strategies

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Abstract

Malignant neoplasms of the appendix are rare and represent less than 1% of gastrointestinal cancers. Goblet cell carcinoids (GCC) tumors are a distinctive group of heterogeneous appendiceal neoplasm that exhibit unique clinical and pathologic features. This review focuses on GCC of the appendix. English language articles from PubMed/Medline and Embase was searched were collected using the phrases "goblet cell carcinoids, adenocarcinoids, crypt cell carcinoma, mixed carcinoid - adenocarcinoma and amphicrine tumors." The current diagnostic procedures, pathogenesis, possible signaling mechanisms and mutations are presented. Treatment options for this neoplasm are reviewed and summarized, although evidence-based data are lacking. Figures, tables and schematic diagram to illustrate pathways are included in the review. Perspectives for future research are presented. Goblet cell carcinoids are a separate entity from carcinoids (NETs) and adenocarcinoma. The prognoses of GCC lays intermediate between appendicular NETs and primary appendiceal adenocarcinoma. The pathogenesis is unclear however the tumor likely arises from

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