

Metastatic Neuroectodermal Tumors of the Gastrointestinal Tract

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Introduction

GNET (malignant gastrointestinal neuroectodermal tumour) is a very rare primary mesenchymal malignancy of the gastrointestinal tract, also known as clear cell sarcoma-like tumour of the gastrointestinal tract (CCSLTGT) or "osteoclast-rich tumour of the gastrointestinal tract with features resembling clear cell sarcoma (CCS) of soft parts." When their 16-case series supported separating GNET from CCS as a distinct tumour entity rather than a variation, Stockman et al. created the term GNET in 2012. As of December 2021, just 111 cases had been recorded, offering a challenge to the limited clinical, prognostic, tumour stage, therapy, and management data available. GNET is also frequently misdiagnosed and treated wrongly due to its rarity and resemblance to other tumours [1-3].

Description

A Caucasian man with stomach cramps, non-bloody diarrhoea, and weight loss presented. Except for a suspected history of small fibre neuropathy shown as intermittent burning sensations in both feet, he was generally healthy, and nerve conduction testing was normal.

A near-obstructing erythematous friable lesion in the sigmoid colon was discovered during colonoscopy. Malignant spindle cells with sporadic pseudoinclusions, moderate cytoplasm, and no evident mitotic patterns were discovered in a biopsy. Calretinin, melanA, HMB-45, chromogranin A, and synaptophysin immunohistochemistry (IHC) staining were positive, while calretinin, melanA, HMB-45, chromogranin A, and synaptophysin immunohistochemistry (IHC) staining were negative.

All stage imaging, including Computed Tomography (CT) and Magnetic Resonance Imaging (MRI), came back negative (MRI). A sigmoid resection was performed laparoscopically. There was no free fluid in the abdomen. A hard tumour of 3-4 cm in length with serosa puckering was detected on the sigmoid colon. On the pelvic peritoneum anteriorly and in the lower left quadrant, there were also modest 1-2 mm flat and soft white deposits. A sigmoid resection and ligation of the low inferior mesenteric artery were performed laparoscopically. Biopsies were taken of a representative sample of the peritoneal deposits.

A 2.2 cm epithelioid and spindle tumour penetrating the full thickness of the bowel from the ulcerated mucosa to the serosal surface was discovered on

pathology. The levels of cellularity ranged from low to high. There was no sign of necrosis on the body. There was no lymphovascular or perineural invasion. Only one out of every 10 high-power fields had a mitotic figure. The tumor's stroma was fibrous and myxoid. Patterns were found on IHC that were similar to those found in the prior biopsy. In most locations, Ki-67 was less than 1%, while extremely small foci showed a greater proliferation rate, though still less than 5% overall. The original tumour had a clean margin. All of the lymph nodes were found to be negative. On the other hand, a peritoneal biopsy revealed.

Conclusion

Despite its rarity, GNET may represent a spectrum of disorders with different histomorphology, clinical presentation, and outcome, as well as response to various systemic treatments. GNET's defining feature and molecular driver is the EWSR1 gene rearrangement, yet it is not a conclusive diagnostic criterion. This and previous studies have shown that reliable GNET diagnosis demands a high level of pathological competence. Recognizing pathological and clinical heterogeneity is crucial not only for accurate diagnosis but also for directing optimal clinical therapy [4,5].

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