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Metastatic Malignant Gastrointestinal Neuroectodermal Tumors

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Introduction

Malignant Gastrointestinal Neuroectodermal Tumour (GNET) is an extremely 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." Stockman et al. coined the term GNET in 2012, when their 16-case series supported distinguishing GNET from CCS as a distinct tumour entity rather than a variant. Only 111 cases had been reported as of December 2021, posing a challenge to the limited clinical, prognostic, tumour staging, or treatment and management information available. Furthermore, because of its rarity and similarities to other cancers, GNET is frequently misdiagnosed and treated incorrectly.

Presentation

A Caucasian male presented with a 5-year history of abdominal cramping, non-bloody diarrhoea, and weight loss. He was otherwise healthy, with the exception of a possible history of small fibre neuropathy manifested as intermittent burning sensations in both feet, but nerve conduction studies were normal. In September 2020, a colonoscopy revealed a near-obstructing erythematous friable lesion in the sigmoid colon. A biopsy revealed malignant spindle cells with occasional pseudoinclusions, moderate cytoplasm, and no obvious mitotic figures. S100, SOX-10, and CD68 immunohistochemistry (IHC) staining were positive, while calretinin, melanA, HMB-45, chromogranin A, synaptophysin, CD117, CD34, muscle specific actin, and broad-spectrum keratin (AE1/3) were negative. Ki-67 was very low, less than 1%. All staging imaging was negative, including computed tomography (CT) and magnetic resonance imaging (MRI). In November 2020, a laparoscopic sigmoid resection was performed. In the abdomen, there was no free fluid. On the sigmoid colon, a firm tumour measuring 3–4 cm in length with a puckering of the serosa was discovered. There were also minor 1–2 mm flat and soft white deposits on the pelvic peritoneum anteriorly and in the lower left quadrant. A laparoscopic sigmoid resection was performed, along with a low inferior mesenteric artery ligation.

Pathology revealed a 2.2 cm French grade 2 epithelioid and spindle tumour infiltrating the entire thickness of the bowel from the ulcerated mucosa to the serosal surface. The cellularity ranged from moderate to high. There was no evidence of necrosis. There was no perineural or lymphovascular invasion. The mitotic figure was sparse, with only one in every ten high power fields. The stroma of the tumour was fibrous and myxoid. IHC revealed patterns that were similar to the previous biopsy. Ki-67 was also less than 1% in most areas, but very small foci demonstrated a higher proliferation rate, albeit less than 5% overall. The margin of the primary tumour was clear. The lymph nodes were all negative. The peritoneal biopsy, on the other hand, revealed metastatic disease. Surprisingly, a EWSR1 fusion was not found in this patient's tumour [1-5].

Conclusion

Despite the fact that GNET is an extremely rare type of sarcoma, it may represent a spectrum of diseases with distinct histomorphology, clinical presentation and outcome, as well as response to various systemic therapeutics. The EWSR1 gene rearrangement is the defining feature and molecular driver of GNET, but it is not a definitive diagnostic criterion. This and other reports have demonstrated that accurate diagnosis of GNET necessitates extensive pathological expertise. Recognizing the level of heterogeneity, both pathologically and clinically, is critical not only for correct diagnosis but also for guiding appropriate clinical management.

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