Sarcomatoid carcinoma of the duodenum

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DESCRIPTION
A 69-year-old Caucasian man presented to the gastroenterology clinic with complaints of nausea, abdominal pain and more than 100-pound unintentional weight loss over the past 1 year. He had a history of colon polyps and was overdue for surveillance colonoscopy. He was subsequently scheduled for an outpatient oesophagastroduodenoscopy (EGD) and colonoscopy as initial work-up for his symptoms. One week later, the patient underwent...
endoscopy. EGD showed an ulcerated mass in the second portion of the duodenum (figure 1). Biopsy of the lesion showed features of pleomorphic cells without glandular differentiation (figures 2 and 3). Immunohistochemical studies revealed pleomorphic cells positive for cytokeratin AE1/AE3 (figure 4), CAM 5.2 and vimentin (figure 5), focally positive for CK7 (figure 6), and weakly for CK20. They were negative for neuroendocrine markers chromogranin, synaptophysin, and CD45 which is a lymphoid marker. Additional stains such as discovered on GIST-1 (DOG-1), C-kit, mouse double minute 2 homolog (MDM2), S-100 and ERG were performed, which were negative. The morphological features and the immunohistochemistry findings were consistent with sarcomatoid carcinoma. A second opinion was sought at a nearby tertiary care centre, and the diagnosis was confirmed. Colonoscopy was negative except for mild diverticulosis in the sigmoid colon. Positron emission tomography-CT demonstrated heterogeneous fluorodeoxyglucose (FDG) active mass in the left upper to a midabdominal area, FDG active mass in the distal pancreatic body, multiple FDG active liver metastasis and mildly FDG active sclerotic focus in the T11 vertebral body (figure 7). The patient was offered palliative radiation, but he was unable to start treatment due to fatigue and poor performance status. He eventually opted for home hospice. Sarcomatoid carcinoma displays properties of both epithelial and mesenchymal carcinomas. It is a rare neoplasm of the small intestine with approximately 30 cases reported to date. Immunohistochemical analysis is necessary to establish a diagnosis since H&E stain exam is not conclusive. Vimentin is positive in approximately 90% of intestinal sarcomatoid carcinomas. Neuroendocrine and neuron-specific markers may be positive in some cases; however, in our case, it was negative. Surgical resection is the first choice of therapy for sarcomatoid carcinoma of the small intestine. Neither radiotherapy nor chemotherapy is known to enhance survival. Sarcomatoid carcinoma is a highly malignant tumour with high mortality, with only a few months of survival duration. In our case the patient survived for 2 months since the date of diagnosis.

Contributors JY performed the EGD and wrote the case description. MPR wrote the description, learning points and edited the draft. MR edited the draft. VA edited the draft.

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REFERENCES