Answer

Angiosarcoma

Histopathological examination of the tissue revealed it to be a high-grade, poorly differentiated sarcoma positive for vimentin and CD31. It was 9 cm in greatest dimension, spanning the full thickness of the jejunal wall, with mucosal ulceration and no angiolymphatic invasion. There were some features of gastrointestinal stromal tumors and Ewing sarcoma; however, owing to the strong CD31 positivity, these diagnoses were excluded. Genetic studies for gastrointestinal stromal tumors like KIT tyrosine kinase and platelet-derived growth factor α were negative. Diagnosis of post-transplant lymphoproliferative disorder was excluded, as in situ hybridization for the Epstein-Barr virus was negative.

Angiosarcomas represent 1% to 2% of all sarcomas and most frequently occur in the skin and subcutaneous tissue. The most frequently affected intra-abdominal organs are the spleen and the liver. Primary gastrointestinal manifestation is quite rare and usually occurs in the stomach or small bowel.[1] Exposure to vinyl chloride, Thorotrast, arsenic chemotherapy, trauma, long-standing lymphedema, and radiotherapy have been implicated in its pathogenesis.[2,3] Clinical presentation usually is a combination of nausea, vomiting, abdominal pain, constipation, and gastrointestinal bleeding.[4] Immunosuppression is responsible for increasing the relative risk of malignancies in transplant recipients.[5]

The histological features of angiosarcoma are similar to Ewing sarcoma, lymphoma, and gastrointestinal stromal tumors, among others. Immunohistochemical staining with expression of endothelial markers such as CD31, CD34, and factor VIII–related antigen is necessary to allow definite diagnosis of angiosarcoma.[6] Sarcomas are rare neoplasms with a 1.7% incidence among all transplant recipients presenting with de novo malignancies.[4] Sarcomas in solid organ transplantation appear to have an aggressive pattern, with 62% being high grade and 40% being metastatic at the time of primary diagnosis.[7]

Transplant recipients with newly diagnosed sarcomas should be treated with multimodal therapy owing to increased incidence of high-grade tumors and a higher rate of recurrence. The mainstay of treatment is complete surgical excision, as in our case. Adriamycin-based chemotherapy can be used as an adjunct. There is also a role of switching to rapamycin-based immunosuppression.[7]

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