Metastatic Angiosarcoma and Kasabach-Merritt Syndrome

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Metastatic angiosarcoma and Kasabach-Merritt syndrome

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Abstract

Angiosarcomas are exceedingly rare tumors that are often difficult to diagnose. Exceptionally unusual is the presentation of these tumors with Kasabach-Merritt Syndrome, a curious form of intratumoral coagulation that can be impossible to distinguish from intravascular coagulation, which is more common. Instant recognition of this clinical association can help making a prompt diagnosis and timely initiation of therapy.

Case Report

A 75-year-old woman presented to her community hospital with headaches and severe thrombocytopenia. A provisional diagnosis of thrombotic thrombocytopenic purpura was made and she was treated with steroids, plasma exchange, then intravenous immunoglobulin and rituximab, with no improvement in platelet counts. She was then referred to us for further management. Upon evaluation, she was noted to have scattered petechiae and a purplish right forehead scalp swelling that she reported had been growing over the previous several months and was initially thought to be a hematoma. She otherwise appeared healthy. Her laboratory findings were remarkable for a platelet count of 18,000/µL (normal 189-377 k/µL), a hemoglobin of 7.9 (normal 12.0-14.1 g/dL), and a white blood cell count of 24,000 (normal 4.1-10.8 K/UL). Direct Coomb’s test was negative. Prothrombin time was 14.7 seconds (normal <11.6), and activated partial thromboplastin time was 30 seconds (normal <11.6), and activated partial thromboplastin time was 30 seconds (normal <11.6). Fibrinogen was 67 mg/dL (normal 150-450 mg/dL), D-dimer 33.97 mg/L (normal <0.50 mg/L FEU) and LDH was 1129 (normal 122-220 u/L); findings most consistent with a consumptive coagulopathy. Computerized tomography (B and C).
from the disseminated intratumoral coagulation that characterizes Kasabach-Merritt syndrome. Recognizing the association between cystic vascular lesions in an elderly patient, consumptive coagulopathy, and the lack of heparin effect on platelet counts may hint at the pathophysiology of the underlying coagulopathy and help make the diagnosis earlier.

References