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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 count. 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/UL (normal 189-377 k/UL), 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 <29). 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 revealed that the scalp lesion was eroding into the skull and extending into the brain parenchyma (Figure 1A), with extensive hepatosplenic involvement with variable sized complex cysts that was photopenic on positron emission tomography (Figure 1B,C).

In addition to transfusion support, a low dose heparin infusion was attempted with no improvement in platelet counts. Biopsy of the scalp lesion was deemed too risky and fine needle aspiration of the liver was nondiagnostic. Unfortunately, the patient experienced sudden deterioration in mental status while hospitalized and expired due to intracranial bleeding from the scalp mass. Permission for an autopsy was granted. The post-mortem examination revealed numerous foci of hemorrhagic necrotic tumors present throughout the liver and spleen (Figure 2A,B) with a single additional focus of tumor extending from the scalp to the intracranial space (Figure 2C,D). Microscopically, the tumor had irregularly anastomosing vascular channels with a dissecting growth pattern lined by high grade malignant endothelial cells with positive membrane staining for CD31 and 34 (Figure 3), findings consistent with high-grade angiosarcoma, likely originating in the scalp.

Discussion and Conclusions

Angiosarcomas are exceedingly rare tumors, representing less than 1% of all sarcomas combined. They are most frequently of skin and soft tissue origin in the head and neck region, but may also arise from other organs such as the liver, spleen, heart, and breast. In particular, angiosarcomas are known to be associated with radiation, chronic lymphedema after breast surgery (Stewart-Treves syndrome), and most recently a possible association with silicone breast implants has also been described. Angiosarcomas exhibit a high propensity for metastatic multifocal disease and carry a poor prognosis. Establishing the diagnosis can be extremely difficult, particularly when angiosarcoma involves visceral organs and when it presents with unusual clinical manifestations.

Somewhat peculiar to angiosarcoma is that it may present with Kasabach-Merritt syndrome, a consumptive coagulopathy originally described in association with large benign hemangiomas in infants. Pathogenesis is presumably related to platelet trapping within these highly vascular lesions with secondary consumption of clotting factors and resultant bleeding. The association of Kasabach-Merritt syndrome with angiosarcoma has been largely reported in the dermatology literature, curiously because of the particular association with scalp angiosarcomas in elderly patients.

Disseminated intravascular coagulation, a more commonly encountered problem in cancer patients, may be difficult to distinguish...
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**