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Adult Kasabach-Merritt Syndrome: A Case Report and Review of Literature

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Abstract

The most common benign liver tumors are cavernous Hemangioma. They can reach enormous sizes and cause varied complications. Kasabach-Merritt syndrome is a constellation of a hemangioma inclusive of endothelial abnormalities, which lead to a consumptive coagulopathy and subsequent thrombocytopenia. Although rare, serious complications are present yielding a mortality rate of 10% to 37%. 80% of cases occur in the first year of life with 0.3% of infants with hemangiomas affected. The treatment aims to control the coagulopathy and thrombocytopenia as well as eradicate the hemangioma. Different treatment approaches are present inclusive of surgical and non-surgical techniques of which systemic corticosteroids, irradiation and immune-suppressors are an option. Surgery is limited to cases of symptomatic and complicated presentations. Although not simple, enucleation is curative usually. Here, we present a 53-year-old man with giant hemangiomas causing KMS.

Keywords: Hemangiomas; Liver; Kasabach-Merritt Syndrome (KMS)

Introduction

Being first diagnosed in 1940 by Haig Haigouni Kasabach and Katharine Krom Merritt, who had been taking care of an infant with a giant hemangioma and thrombocytopenia purpura, Kasabach Merritt Syndrome (KMS) is a rare, but serious constellation of a hemangioma within which exist endothelial malformations that cause a consumptive coagulopathy leading to a thrombocytopenia [1-3]. Such lesions exist typically superficial and solitary in a male infant, which represent 80% of cases while adult KMS exists in 20% of cases where the hemangioma exists in internal organs such as the liver [4]. It is well documented that the most common benign tumors of the liver are cavernous hemangiomas. Those larger than 4 cm are termed giant hemangiomas. With the advantage of reduced blood loss and virtually ability to preserve all viable liver tissue, surgical resection is the typical usually curative treatment approach, which is preserved for complicated and symptomatic cases [5-7]. Here, we present a 53-year-old man with giant hemangiomas causing KMS.

Case Presentation

A 53-year-old Iraqi man, previously diagnosed with ITP (Idiopathic Thrombocytopenic Purpura) in 1990 treated irregularly by prednisone without sustained response, and DVT (Deep Vein Thrombosis) in 1992 treated by anti-vitamin K, presented in April 2018 for second opinion about his asymptomatic persistent thrombocytopenia. For 5 months before presentation, despite treatment with prednisone (50 mg) and azathioprine (100 mg), the patient had platelet count ranging between 5000/mm^3 to 20000/mm^3 with no clinical symptoms.

The patient is a non-smoker, non-alcoholic with negative history of diabetes mellitus, hypertension and coronary artery disease. The clinical exam was normal with no signs of bleeding. The patient was afebrile, conscious, oriented and cooperative.

At presentation, the complete blood count revealed 27,000 platelets/mm^3, normal white blood cell count (6700/mm^3) and hematocrit of 41%. Bone marrow aspirate showed hyper cellularity and normality in the three cell lineages. The physical examination was normal.

An abdominal US performed at presentation showed a very heterogeneous poorly limited hypo-echoic nodule formation diffuse through the liver (Figure 1). Abdominal MRI (Figure 2, 3) revealed two large liver masses both showing peripheral enhancement after gadolinium injection. The first is a well-defined 13.5 cm mass of right hepatic lobe involving segments VII and VI. The 2nd is a sub-capsular mass of segment VIII and IV of 10 cm size with adjacent smaller 2.5 cm nodular lesions of

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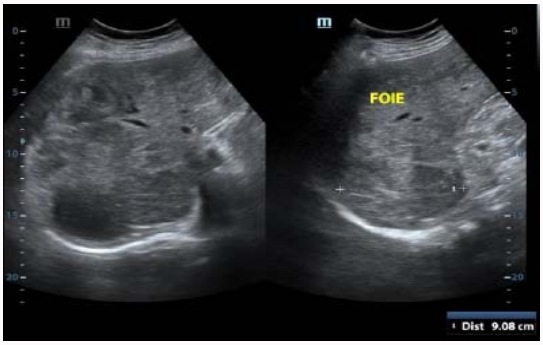

Figure 1: Ultrasound of the liver.

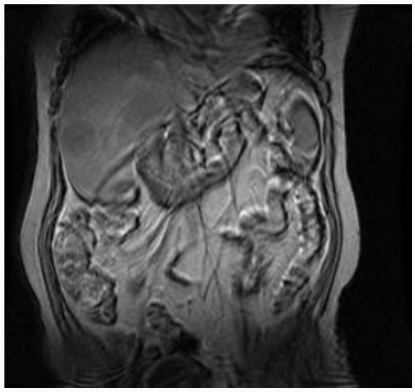

Figure 2: Frontal view of abdominal MRI.

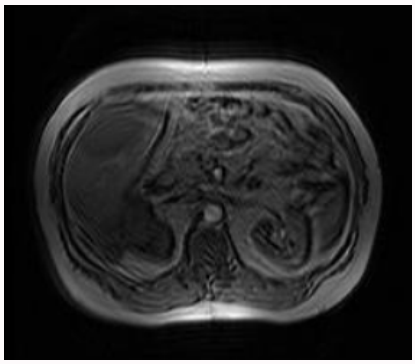

Figure 3: Transverse view of Abdominal MRI.

segment IV and III.

As part of complete laboratory workup, anticardiolipin antibodies were quantified and revealed positive suggestive of antiphospholipid syndrome while liver findings are typically suggestive of adult KMS. In addition, PCR was done for CMV nucleic acids that have proved 2.96-log copies/ml. No treatment was held as the patient returned back to Iraq with the diagnosis of KMS and recommendation for CMV treatment (probably secondary the ongoing immunosuppressive treatment), and consideration of surgical approach.

Discussion

Kasabach-Merritt results in a consumptive coagulopathy [1,3] secondary to platelet trapping and aggregation within a specific type of hemangioma, and can have a high mortality rate ranging between 1% to 37% [8]. In infants, the hemangiomas are typical confined to the skin, yet they can present in retroperitoneal organs such as the mediastinum, the pelvis, visceral organs, or the mesentery. This patient presented with a giant liver hemangioma and asymptomatic thrombocytopenia. Although anticardiolipin antibodies proved positive giving indication of antiphospholipid syndrome, without repeated laboratory testing of anti-b2 glycoprotein-I antibody of IgG and/or IgM isotype and/or IgG or IgM subtype of anticardiolipin antibodies, it is recommended to avoid APLS diagnosis. The positive result of these antibodies does not certify ultimate diagnosis of APLS. In the presence of a giant hemangioma in the liver correlating with laboratory result of thrombocytopenia, and absence of other clinical symptoms including rash, skin patches, repeated episodes of DVT and PE, Kasabach Merritt syndrome is more probable. In addition, treatment with prednisone, anti-vitamin-k and azathioprine has not proved effective which verifies suggestions of KMS over APLS.

Management of this case of KMS mandates non-surgical approach. First line treatments are vincristine and corticosteroid therapy. The angiogenic character of Kasabach-Merritt syndrome indicates that chemotherapy is a logical treatment which has been proven effective in a multicenter study in the United States [9]. Enjolras et al. have reported that several steroid non responders show dramatic response to vincristine [10].

Trails with variable results have been made for alternative therapies, including interferon α-2a and 2b [11], radiation therapy and chemotherapeutic agents such as actinomycin. The most promising recent option available for treatment of infantile hemangiomas is oral solutions of propranolol [12].

Surgical approach remains a mainstay in the treatment of symptomatic and complicated KMS cases [6]. This approach is recommended for single cutaneous lesions or multiple lesions in the spleen (splenectomy) or liver (wedge resection/hepatectomy) [3,13,14]. It is proved to be the only curative treatment of KMS. In this patient, resection has two major advantages: Minimal blood loss and preservation of virtually all-viable liver tissue [7].

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