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Case Report

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A Rare Case of Kasabach-Merritt Syndrome Presenting with Infantile Hemangioma

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1. Introduction

Infantile hemangioma is a proliferative hamartoma of the vascular endothelium, with factors such as GLUT-1 receptors, increased vascular growth factors, and inappropriate distribution of chorionic villus cells during fetal life playing a role in its etiology and pathogenesis [1]. The association of hemangioma with thrombocytopenia and hypofibrinogenemia was first described in 1940 by Kasabach and Merritt, who reported a baby with a giant capillary hemangioma and thrombocytopenic purpura [1,2]. The classic triad of Kasabach-Merritt Syndrome consists of hemangioma, thrombocytopenia, and hypofibrinogenemia [2]. A case of Kasabach-Merritt Syndrome was presented, which was identified on antenatal ultrasound as a liver hemangioma and followed up during the postnatal period.

2. Case Report

A 39-week-old girl, born c/s to a 29-year-old mother in her fourth pregnancy (second living), had an APGAR score of 9/10 at both 1 and 5 minutes. The infant was admitted to the neonatal intensive care unit for monitoring and treatment due to a liver hemangioma detected at 7 months gestation via antenatal ultrasound. On physical examination, her body weight was 2945 grams, head circumference 33 cm, and there was palpable hepatomegaly of 3 cm below the costal margin. No widespread petechiae or ecchymoses were observed upon examination of the entire body. The mother's blood count was normal. The infant had thrombocytopenia (102,000 µL). To assess the development of disseminated intravascular coagulation (DIC), tests for fibrinogen, PT, aPTT, and INR were requested. Results were as follows: fibrinogen 261 mg/dL, PT 10.60 s, aPTT 37.1 s, INR 1.12. An abdominal ultrasound revealed a hyper-echoic lesion in the right lobe of the liver, consistent with a 50x45 mm hemangioma in segments 6-7.

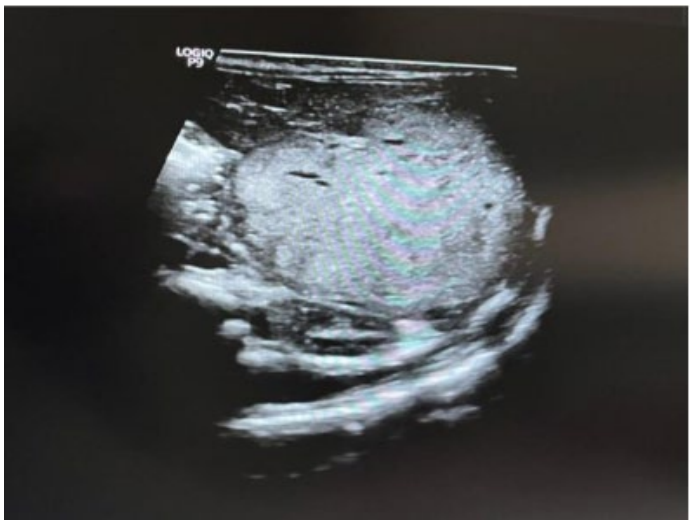

Figure 1: A Hyper-Echoic Lesion that Could be Consistent with A 50x45 mm Hemangioma

In our case, intermittent testing was planned for DIC and bleeding in surrounding tissues, and propranolol (2 mg/kg/day) was initiated. AFP was intermittently tested to differentiate between hemangioma and hepatoblastoma. Follow-up results showed platelet counts of 97,000 µL, 159,000 µL, and 248,000 µL, and coagulation parameters were: fibrinogen 197 mg/dL, PT 9.56 s, aPTT 29.5 s, INR 1.02. The patient was discharged after being deemed suitable for follow-up by pediatric hematology. During follow-up by pediatric hematology, MRI and abdominal ultrasound showed that the hemangioma had grown. An MRI revealed a lesion approximately 5x6.5 cm in size, covering the entire right lobe of the liver. It appeared heterogeneously hyperintense on T2-weighted sequences, heterogeneously hypointense on T1-weighted sequences, and showed lobulated contour with peripheral enhancement after contrast administration, characteristics that could be consistent with hepatoblastoma.

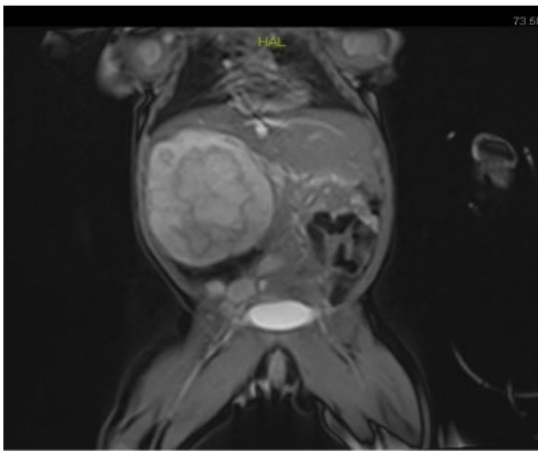

Figure 2: 5x6.5 cm A Lesion that May Be Compatible with Hepatobulbar

AFP decreased naturally from 110,775 µg/L to 30,797 µg/L, distancing the diagnosis from hepatoblastoma. Upon observing growth in the hemangioma, further follow-up and treatment were initiated. During follow-up, the patient had a hemoglobin level of 6.8 g/dL, suggesting possible intralesional bleeding. She was transfused with 10 mL/kg of blood, increasing her hemoglobin to 9 g/dL. Oral iron was added to the treatment plan, and MRI and portal Doppler ultrasound were scheduled. The portal Doppler ultrasound showed the liver size increased to 92 mm. The parenchymal echo was homogeneous. The left lobe of the liver extended variably towards the anterior of the spleen. A lobulated, hypoechoic, heterogenous solid lesion with cystic degenerative areas and possible calcifications, measuring 75x64x86 mm and nearly filling the right lobe of the liver, was observed. The patient was referred to Uludağ University for biopsy by the hematology-oncology department. The pediatric tumor board recommended continuing propranolol at 2 mg/kg/day, initiating prednisolone at 2 mg/kg/day for 5-7 days. Malignancy was not suspected based on ultrasound and MRI images. AFP decreased further from 30,797 µg/L to 777 µg/L, supporting the diagnosis of hemangioma. Due to the decrease in AFP and the presence of thrombocytopenia, the biopsy was postponed. The patient was admitted to the ward, and intravenous prednisolone treatment was completed in 7 days. A follow-up abdominal ultrasound revealed a slightly lobulated, solid lesion in segments 7-6-5, measuring approximately 5.5x4 cm, with coarse calcifications, and a slightly hyperechoic appearance compared to the surrounding parenchyma. The hemangioma decreased in size upon follow-up.

3. Discussion

Kasabach-Merritt syndrome is a combination of a rapidly growing hemangioma, thrombocytopenia, and acute or chronic consumptive coagulopathy [3]. Clinical manifestations usually appear in early infancy but can also present later [3]. The associated thrombocytopenia can lead to bleeding, ecchymosis, petechiae, and rapid growth of the hemangioma [3]. Severe anemia may occur due to bleeding or microangiopathic hemolysis [3,4]. The cause of thrombocytopenia is either sequestration or increased destruction of platelets within the hemangioma [4]. Treatment involves managing thrombocytopenia, anemia, and consumptive coagulopathy through the administration of platelets, red blood cells, and fresh frozen plasma [4]. The goal of treatment is to prevent significant morbidity or mortality or to induce tumor involution in response to life-threatening events [4,5]. Surgical excision is curative but is not suitable for most lesions [5]. Historically, the first-line treatment has been high-dose systemic corticosteroids. However, two-thirds of lesions will not respond to corticosteroids or will rapidly recur when treatment is discontinued [5]. Various alternative treatments have been tried with variable results, including interferon α−2a and α−2b, radiation therapy, and chemotherapy agents such as vincristine and actinomycin [6]. In our case, corticosteroids were the initial treatment of choice. Studies have shown that corticosteroids have been used both as a single therapy and in combination with other treatments (such as radiation therapy) at high doses (2-3 mg/kg/day of prednisone) to achieve a response. In our case, prednisolone was administered at the doses specified in the studies (2 mg/kg/day) [6]. After a total of 1 week of treatment, the patient's hematological parameters returned to normal, and there was a dramatic reduction in the size of the lesion.

4. Conclusion

If Kasabach-Merritt syndrome (KMS) and disseminated intravascular coagulation (DIC) develop in vascular tumors, they can increase the risk of mortality and morbidity by causing bleeding. Based on this case, it is important to monitor patients diagnosed with KMS in the antenatal period for DIC in the postnatal period as well. We concluded that systemic corticosteroids are an effective and safe treatment for this newborn, and hepatoblastoma should be considered in the differential diagnosis.

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