Diffuse cavernous hemangioma of the spleen with Kasabach-Merritt syndrome misdiagnosed as idiopathic thrombocytopenia in a child

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Background: **Most cavernous hemangiomas in the spleen are small lesions that are found incidentally and patients usually present with no symptoms. Imaging is able to detect the lesions that are considered as diagnostic evidence. But some patients with diffuse cavernous hemangioma may present with anemia, thrombocytopenia, coagulopathy and bleeding, which might be misdiagnosed as idiopathic thrombocytopenia with disseminated intravascular coagulation (DIC). Splenectomy is the most effective therapy for diffuse cavernous hemangiomas with symptoms.**

Methods: **The history, imaging results, pathologic findings, diagnosis and treatment of a 34-month-old boy with severe petechiae were reviewed.**

Results: **The boy was diagnosed as having refractory idiopathic thrombocytopenia (ITP) because of low platelet count and bleeding at a local hospital. He had no response to a full-dose of corticosteroid and a high-dose of immunoglobulin (2 g/kg). Huge splenomegaly and DIC were found after 7 months. Diffuse cavernous hemangioma of the spleen was highly suspected, but it was not confirmed by B ultrasound, enhanced CT or MRI. DIC and bleeding were solved by low molecular weight heparin, supplement of fibrinogen and prothrombin complex. A diffuse cavernous hemangioma involving the whole spleen was confirmed pathologically following a successful splenectomy. The boy recovered completely without any complication after the operation.**

Conclusions: **Diffuse cavernous hemangioma of the spleen should be differentiated from ITP associated with splenomegaly. Radiological and overall physical examination should be emphasized for refractory ITP cases.**

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Key words: **diffuse cavernous hemangioma; idiopathic thrombocytopenia; Kasabach-Merritt syndrome; misdiagnosis**

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Introduction

Most cavernous hemangiomas of the spleen are small lesions that are found incidentally and patients usually have no symptoms. The natural course of cavernous hemangiomas is slow, and the symptoms or complications, when present, occur late. Cavernous hemangiomas of the spleen can be large and manifest as a palpable nontender mass in the left upper quadrant. Generalized splenomegaly may be present, but the results of laboratory evaluation are often normal. With anemia, thrombocytopenia, and coagulopathy, cavernous hemangiomas is recognized as Kasabach-Merritt syndrome, which has been reported in patients with diffuse cavernous hemangioma in the spleen. Usually it can be easily confirmed by imaging study. Splenectomy is curative for such patients with symptoms.[1] We report a young boy with a huge spleen, refractory low blood platelet count (BPC), disseminated intravascular coagulation (DIC) but no typical characteristics of cavernous hemangioma shown by CT, MRI or B ultrasound.

Case report

A 34-month-old boy with recurrent petechiae for 7 months and hemorrhage in the left eye for over 1 month was transferred to our hospital. He was diagnosed as having idiopathic thrombocytopenia (ITP) at a local

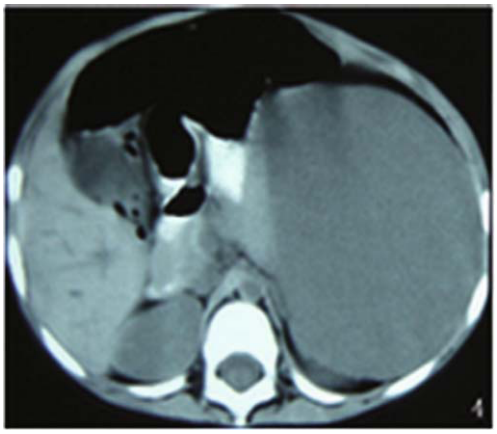

Fig. 1. Unenhanced CT showing only homogeneous hypo-attenuation.

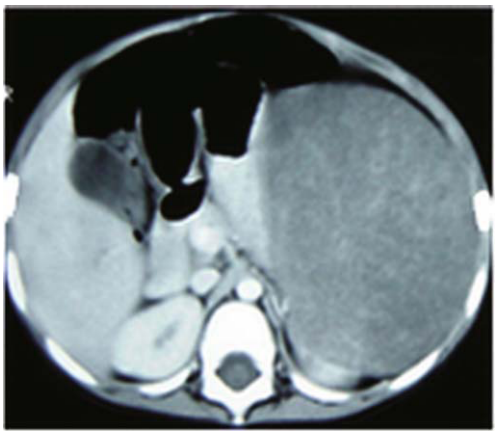

Fig. 2. Enhanced CT showing late- or delayed-phase homogeneous contrast enhancement and no discrete mottled areas of heterogeneous attenuation.

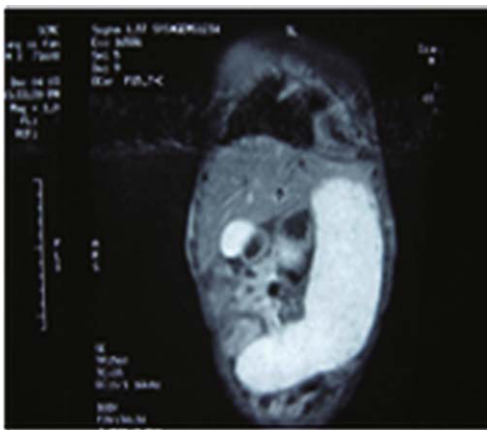

Fig. 3. MRI of coronal T1-weighted image after contrast showing a huge spleen in the pelvis with homogeneous enhancement.

Fig. 4. The spleen was weighted 1000 g. The cut-surface of the whole spleen was reddish, soft, and spongy.

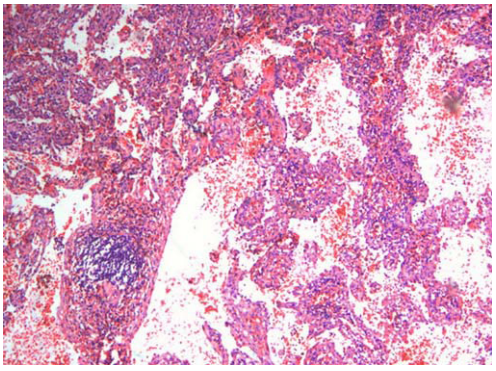

Fig. 5. Under low magnification, the tumor was composed of the vessels arranged in a diffuse pattern.

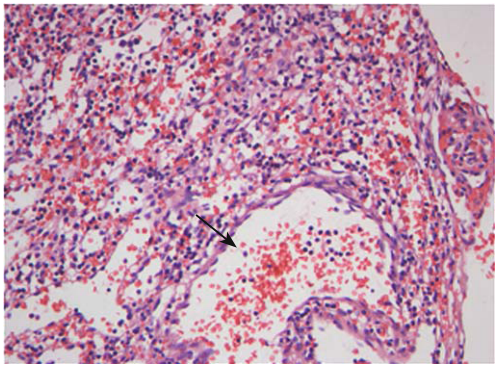

Fig. 6. Under high magnification, the vessels were dilated, blood-filled and lined by flattened endothelium (arrow).

hospital 6 months ago. Laboratory test there revealed: WBC 40×10^9/L, Hb 102 g/L, and BPC 20×10^9/L; bone marrow examination suggested ITP. A full-dose of corticosteroid and a high-dose of immuno-globulin (2 g/kg) were given at the local hospital, but with no response. There was always a bleeding tendency, and platelet count was persistently kept at a low level of $5\text{-}20\times10^9$/L. The blood count at our hospital was as follows: WBC 1.4×10^9/L, Hb 82 g/L, and BPC 20×10^9/L. Physical examination showed blood pressure of 90/60

mmHg. Severe petechiae could be found at the head, face, especially at the left orbit and conjunctiva of the left eye as well as in the body, lower extremities. Heart and lung examination revealed nothing abnormal. Liver was palpated 1.5 cm below the right costal margin and a huge splenomegaly, which had never been mentioned at the local hospital, was found 12 cm below the left costal margin, and occupied almost the whole abdomen and pelvis. The results of the whole set of DIC assay were as follows: prothrombin time (PT) 57.4 seconds, activated partial thromboplastin time (APTT) 90.6 seconds, fibrinogen (FIB) <0.03 g/L, and D-D Dimer 0.3 mg/L. The smear slide of bone marrow taken from the local hospital showed hypercellularity with megakaryocytes, including 9 promegakaryocytes, 92 granulotypes of megakaryocyte, and 8 megakaryocytes with platelet formation. No other abnormalities were found. At the same time, the huge splenomegaly was confirmed at our hospital by B ultrasound examination but no signs of diffuse cavernous hemangioma of the spleen or liver were present. This was re-confirmed by enhanced CT and MRI. On un-enhanced CT scans, diffuse cavernous hemangiomas should appear as well-margined hypo-attenuation or iso-attenuation masses, but in this case, there was only homogeneous hypo-attenuation (Fig. 1). On enhanced CT scans, diffuse cavernous hemangiomas should have a combination of solid and cystic components. The solid component appears iso-attenuation or hypo-attenuation comparing with normal spleen, with enhancement of only the solid tissue. But in this case, there were only late- or delayed-phase homogeneous contrast enhancement and no discrete mottled areas of heterogeneous attenuation (Fig. 2). The MRI appearance should be mottled hypo- to iso-intense, compared with the normal spleen on T1-weighted images and hyper-intense on T2-weighted images. In this case, coronal T1-weighted image after contrast showed a huge spleen into the pelvis with homogeneous enhancement (Fig. 3).

The patient was treated with low molecular weight hypodermic heparin (flaxiparine) 0.15 ml every 12 hours (0.02 ml/kg/day), with supplement of fibrinogen and prothrombin complex as well as platelets from a single donor. One week later, all the indicators for DIC returned to normal. Then, hypodermic flaxiparin 0.35 ml (0.023 ml/kg/day) was administered continuously for 24 hours with significant improvement of the bleeding tendency, whereas the platelet count still kept at a low level. Because diffuse cavernous hemangioma of the spleen was highly suspected clinically, the patient was indicated for splenectomy. But the operation was refused by his parent because of the possibility of severe infection. The patient was discharged from our hospital with normal results of DIC test but a low platelet count. One month later, he was readmitted with the similar manifestations and laboratory data as the initial admission. The bleeding tendency was more severe. He was treated again with low molecular weight heparin, and fibrinogen and prothrombin complex. When DIC was controlled, he underwent splenectomy successfully with the support of platelet transfusion, fibrinogen and prothrombin complex supplement. Pathological examination found that the spleen was weighted 1000 g and diffusely enlarged. The cut-surface of the spleen was reddish, soft and spongy (Fig. 4). Under low magnification, it was composed of the vessels, which were arranged in a diffuse pattern and the residual follicle can be seen (Fig. 5). Under high magnification, the vessels were dilated, filled with blood and lined by flattened endothelium (Fig. 6).

After splenectomy, the platelet count rapidly returned to normal with disappearance of all the indicators for DIC. Two weeks later, he was discharged from the hospital. The blood cell count and the indicators for DIC remained normal without any bleeding tendency or complication associated with splenectomy when he visited us again after two months.

Discussion

Without careful physical examination at the local hospital, a splenomegaly was neglected for 7 months even it extended into the pelvic cavity. If it was detected early, differentiation diagnosis would be possible. The hypercellularity of megakaryocytes with abnormal maturation in bone marrow was misunderstood as a characteristic pattern only for ITP. Actually, other consumptive thrombocytopenia also had such similar patterns, for instance, over-function due to splenomegaly, which resulted in low platelet count.

Generally speaking, the treatment with corticosteroid can control bleeding tendency in patients with ITP, even when the platelet count still kept at a low level. If the bleeding tendency persisted, other causes should be ruled out. Diffuse cavernous hemangioma inside the body should be highly suspected when the patient was complicated with consumptive coagulopathy. Liver, spleen, mediastinum and bone are the most common places for diffuse cavernous hemangioma.[2-4]

B ultrasound, contrast CT or MRI demonstrate no characteristic patterns for diffuse cavernous hemangioma of the spleen in rare cases. It is difficult for us to diagnose diffuse cavernous hemangioma when lacking imaging characteristics or with only homogenized diffuse congestive splenomegaly. Apparently, imaging study (B ultrasound and enhanced CT) is not sensitive enough to detect all diffuse

cavernous hemangioma of the spleen.[5]

Low molecular weight heparin combined with supplement of the associated coagulation factors could effectively control the consumptive coagulopathy complicated by diffuse cavernous hemangioma.[6-11] But if the tumor is not removed, the consumptive coagulopathy will recur.[12]

Splenectomy is indicated for children with diffuse cavernous hemangioma of the spleen complicated with the consumptive coagulopathy, even in very young children with possible complications of severe infection.[1,13,14] Lethal hemorrhage is more likely than lethal infection if consumptive coagulopathy could not be effectively controlled. Consumptive coagulopathy and low platelet count should be controlled before and during splenectomy to reduce the operative risk.

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