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Surgical treatment of kaposiform hemangioendothelioma in the pelvic cavity, bladder and ureter

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ABSTRACT

Kasabach-Merritt phenomenon (KMP) is a rare potentially life-threatening consumptive coagulopathy characterized by thrombocytopenia and hypofibrinogenemia occurring associated with the vascular tumors kaposiform hemangioendothelioma (KHE) and tufted angioma (TA). KHE in the specific sites will also cause some non-hemorrhagic complications. We report a two-month old female infant with KHE in her pelvic cavity, bladder and ureter who responded well to surgical excision.

1. Introduction

Kasabach-Merritt phenomenon (KMP) is specific to KHE and TA, occurring in 70% of KHE and less with TA [1]. There are many treatments for KHE, such as steroids, cytotoxic agents, interferon, vincristine, interferon, antiplatelet agents, propranolol, Sirolimus, resection, and embolization [2–4]. The treatment of KHE need to be individualize in clinical practice. KHE occurs frequently in the trunk, extremities, and maxillofacial regions [5]. We report a rare case of KHE in the pelvis, bladder, and ureter, resulting in double hydronephrosis and ureteral stenosis complications. The patient had used hormone and interferon and made a desirable result. Although the platelet maintain a normal level, persistent KHE leads to serious complications of urinary system. We use surgical resection of the tumor to remove the compression factors and reduce urinary complications (see Figs. 1–4 and Graph 1).

2. Case report

There was a 2 cm × 3 cm mass in the abdominal wall after the birth of the female infant. After 19 days, the abdominal wall and perineal area appear purple ecchymosis, at the same time the mass increased rapidly to approximately 10 cm × 8 cm. The infant underwent routine blood tests to show thrombocytopenia (5 × 109/L) and transfused platelets urgently. Subsequently, the infant was transferred to the higher lever hospital for color Doppler examination and CT examination which showed bilateral hydronephrosis. A little mass was taken for pathological examination and double nephrostomy was performed. Pathological report showed that the mass was KHE. With methylprednisolone, interferon and captopril treatment, the blood platelet gradually rose to 220 × 10^9/L in a month. The infant urinated 4 times through the bladder a week. Subsequently, the infant came to the Department of hemangioma &Vascular Malformation for surgical treatment. During the operation, we could see that the tumor invaded the abdominal wall muscles, anterior bladder wall, the lateral wall, bilateral ureter orifice, pubic symphysis posterior. The tumor was stiff and tightly adherent. The elasticity of the anterior bladder wall decreased significantly. The operation lasted 4 h, with a hemorrhage of 275 ml, accompanied by transfusion of plasma, erythrocytes, and cryoprecipitate. After operation, the infant was transferred to pediatric intensive care unit to treatment. Platelet and coagulation indexes were normal. All vital signs of the patient were good and hormone gradually decreased until withdrawal in two weeks. Then methylene blue test showed positive, and angiography T tube showed distal ureteral stenosis which need to be treatment after two months by pediatric surgeon. The frequency of urination and urine volume increased obviously through the bladder a day. At present, the patient has been discharged.

3. Conclusion

Kaposiform hemangioendothelioma (KHE), is an intermediate/borderline vascular neoplasm between a hemangioma and a malignant angiosarcoma. It is a locally aggressive, rarely metastatic neoplasm,

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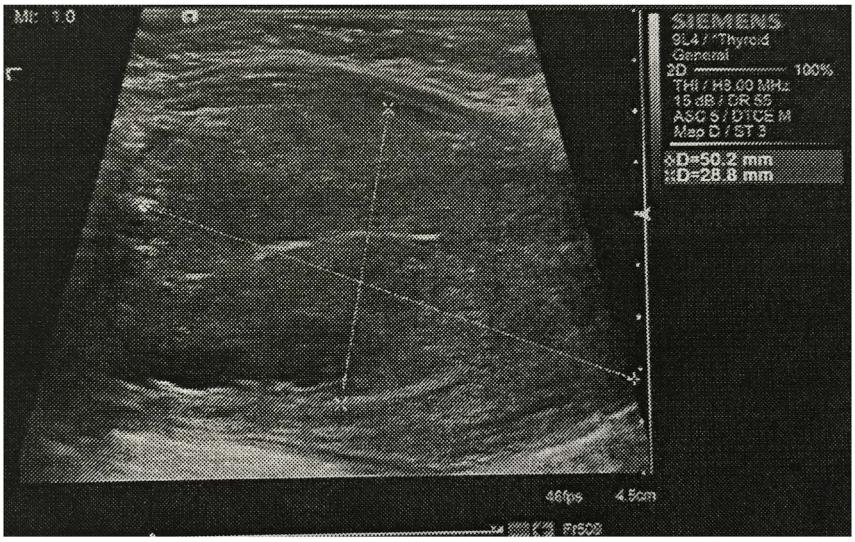

Fig. 1. Ultrasonographic findings of KHE. Ultrasound indicated that there was no clear boundary between pelvic cavity, pelvic floor solid mass and bladder wall.

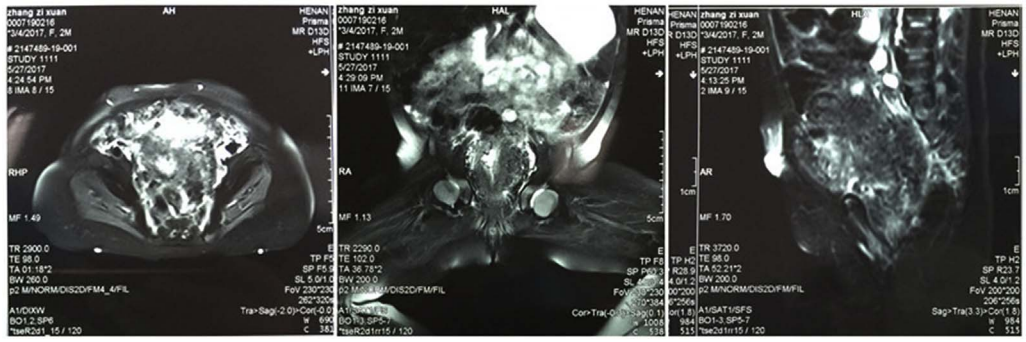

Fig. 2. Multi-phase axial computerized tomography images of KHE. MRI showing the presence of voluminous masses in the pelvic cavity.

does not have a tendency for spontaneous regression and has characteristic histopathological features [6]. Basic diagnostic criteria of KHE are hemangioma and low platelet. Ultrasonography and MRI reveal the size, appearance, and layers of hemangioma, as well as its relationship with peripheral blood vessels, and distinguish hemangioma from vascular malformations. The management of KHE is very challenging because of its rarity and unstandardized therapeutic strategies. The KHE always may requires a combination of treatments. Drolet et al. considered total resection was often not a viable option given the high risk of morbidity and mortality, and due to their infiltrative nature and the invading of large neurovascular structures, surgical resection was not considered to be a primary treatment option [7]. Although it is not recommended, it is a better option for rapid removal of primary disease and reduction of complications in the special parts. Complete surgical resection offers the most definitive cure for small, localized tumors. So prompt diagnosis and initiation of therapy are critical in KMP, which, if not treated, is associated with high mortality [8]. For our patient with KHE, we elected surgery for the three reasons. Drug treatment for a long time, tumor's proliferative activity, compression and invasion, causing irreversible damage to the urinary system until a life. So surgical resection may be more effective with less side effects and is usually curative with a complete reversal of coagulopathy and removal of pathogeny. Each treatment has its own advantages and disadvantages, and effective feasible method which be good at by doctor for the patient can significantly improve the cure rate.

Conflicts of interest

We declare that we have no financial and personal relationships with other people or organizations that can inappropriately influence our work, there is no professional or other personal interest of any nature or kind in any product, service and/or company that could be construed as influencing the position presented in, or the review of, the manuscript entitled, “Surgical treatment of kaposiform hemangioendothelioma in pelvic cavity, bladder and ureter: A case study”.