

Surgical treatment of a huge kaposiform hemangioendothelioma in the chest wall: A case study

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Abstract

Kaposiform hemangioendothelioma, a rare vascular pediatric tumor often associated with Kasabach–Merritt phenomenon, is characterized by severe thrombocytopenia and consumptive coagulopathy. Kaposiform hemangioendothelioma is a severe disease and may progress quickly, resulting in a high mortality. However, standard treatment regimens for Kasabach–Merritt phenomenon have not yet been established. We reported here an infant with a large congenital kaposiform hemangioendothelioma in his chest wall who responded extremely well to surgical excision.

Keywords

Kaposiform hemangioendothelioma, Kasabach–Merritt phenomenon, surgical excision, thrombocytopenia, coagulopathy, therapy

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Introduction

Kaposiform hemangioendothelioma (KHE) with Kasabach–Merritt phenomenon (KMP) is characterized by severe thrombocytopenia and consumptive coagulopathy. The treatment of KHE is very challenging and may require a variety of approaches, such as steroids, cytotoxic agents, interferon, vincristine, radiation, resection, and embolization. In this study, we described the medical process of a 48-day-old full-term male infant, with a enormous KHE in his chest wall when he was born and a serious KMP a few days later. The patient did not respond to propranolol or glucocorticoid, however, recovered amazingly after receiving a surgical treatment in our center. The success of this case may remind surgeons to take surgical approach to treat localized KHE, which could also avoid the side effect of drug therapy and residual lesions of KHE to a large extent. However, we still recommend nonsurgical treatment for disseminated KHE to avoid tough recovery.

Case report

A 48-day-old full-term male infant was presented to the Department of Hemangioma & Vascular Malformation, with an enormous violaceous mass extending to the chest wall (Figure 1). The enormous mass was found through color

doppler ultrasound before the baby was born, and the computed tomography (CT) scan taken on admission showed an infiltrating mass ($10.14 \times 11.59 \times 4.15 \text{ cm}^3$) extending from above the umbilicus to the neck, whereas the initial laboratory evaluations did not show any abnormality until a week later. Propranolol was given immediately in the following month, but it did not work on the patient, and the counting of platelets (PLT) decreased significantly day by day. Then, tissue biopsy confirmed that the mass is a KHE. Glucocorticoid (1.5 mg/kg/day for the first day, then 2.0 mg/kg/day for the remaining 4 days) was given for 5 days, but did not show any effect, and PLT transfusion was not effective either. Taken together, all the therapies above given by other medical center did not show any effect toward the KHE.

After the patient was hospitalized in our center, we took a series of laboratory and imaging tests. The blood tests showed severe anemia and thrombocytopenia (Graph 1).

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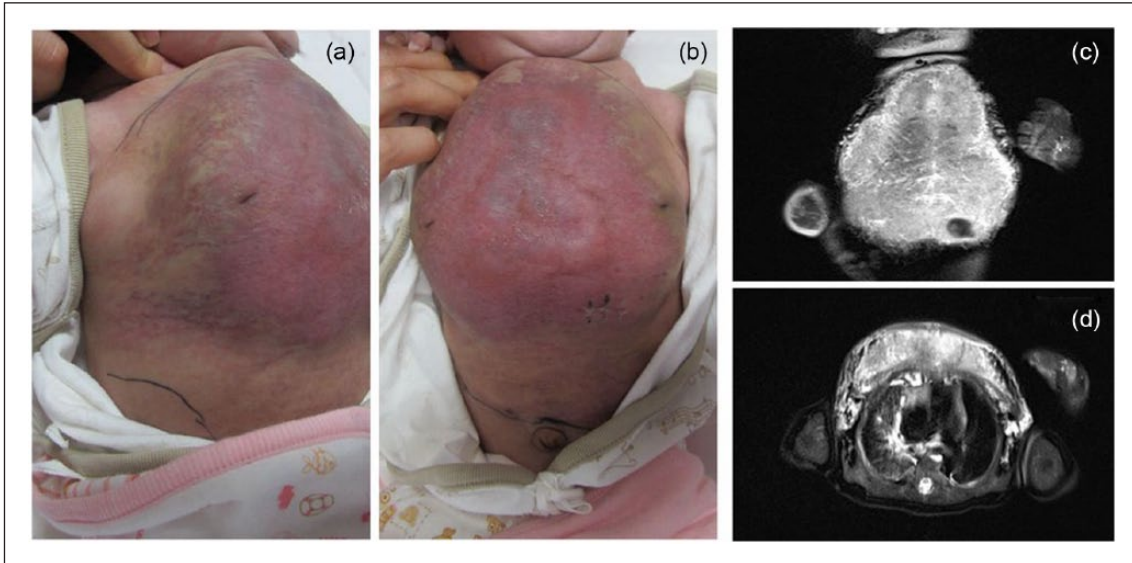


Figure 1. (a, b) A 48-day-old full-term male infant with an enormous violaceous mass extending to the chest wall and (c, d) MRI showing the presence of voluminous masses surrounding the thorax.

Relevant haematological investigations carried out during hospital stay					
Hospital stay	Investigations				Treatment
	Platelet (109/L)	RBC (1012/L)	HGB (g/L)	others	
1st day	17	3.1	94	PT=22.7s (N = 11–17) INR=1.87 (N=0.8–1.5) FIB=0.17g/L (2–4) D-dimmer=10.14µg/ml (N=0–0.5)	prednisone vitamin k1
2nd day	16	3	92		
3rd day	32	3.1	95		
8th day	18	2.6	78		surgery
9th day	84	4.4	131		
13th day	453	4.08	120	PT=13.5s (N = 11–17) INR=1.04 (N=0.8–1.5) FIB=2.89g/L (2–4) D-dimmer=0.8µg/ml (N=0–0.5)	
3rd week	413	4.6	138		
1st month	263	4	119		
38th day	313	3.31	98	PT=17s (N = 11–17) INR=1.31 (N=0.8–1.5) FIB=4.64g/L (2–4) D-dimmer=0.4µg/ml (N=0–0.5)	
52th day (discharge)	265	3.2	99	PT=17.9s (N = 11–17) INR=1.38 (N=0.8–1.5) FIB=2.27g/L (2–4) D-dimmer=0.2µg/ml (N=0–0.5)	

Graph 1. Relevant hematological investigations carried out during hospital stay.

Magnetic resonance imaging (MRI) confirmed the presence of voluminous masses surrounding the thorax (Figure 1). Taken together, these tests confirmed the diagnosis of KMP

of the patient. In our center, the patient was first treated by a high-dose methylprednisolone sodium succinate prednisone (3 mg/kg per day), and vitamin k1 was given for 6 days,

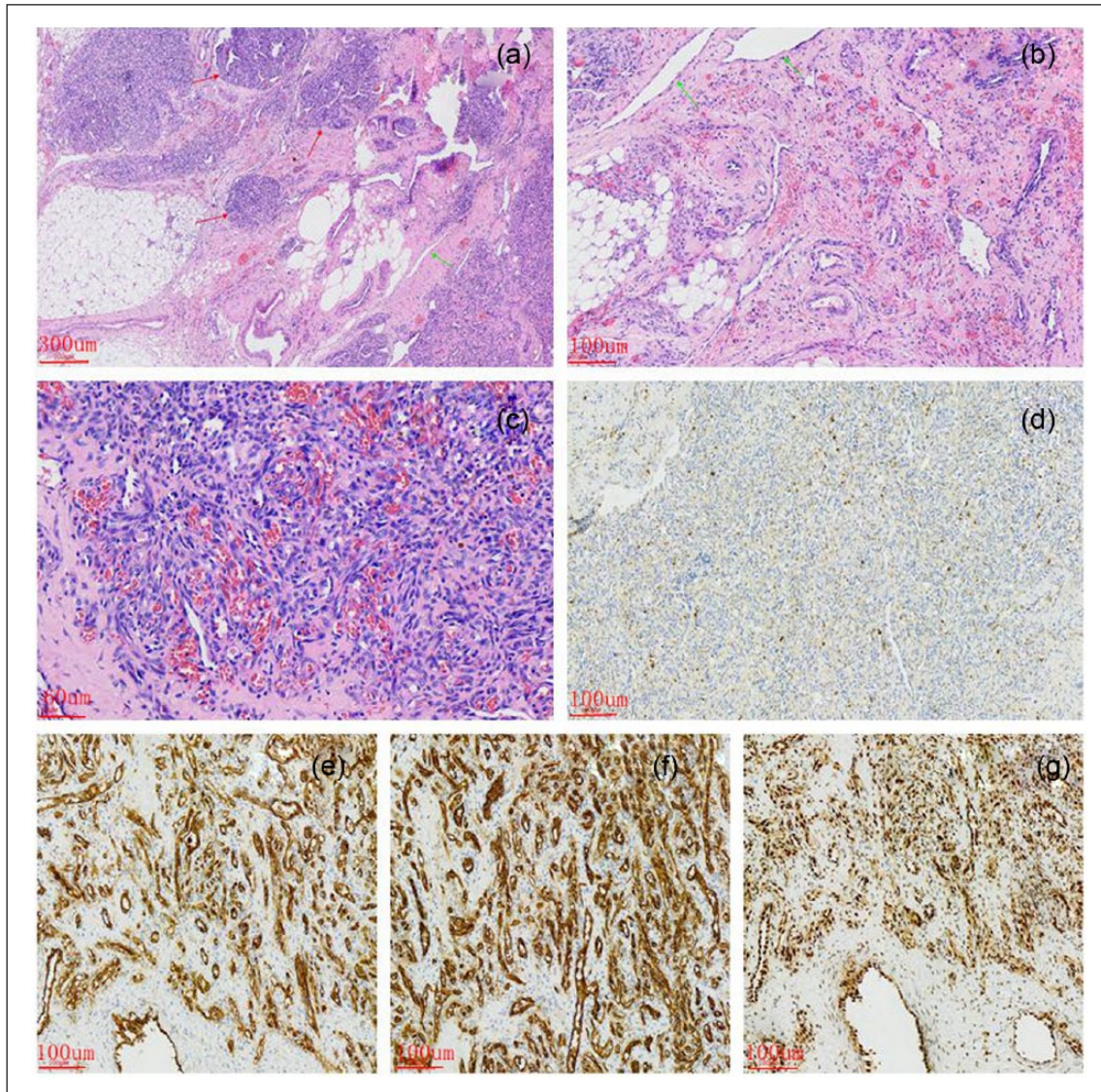


Figure 2. (a) Hematoxylin–eosin (HE) staining $\times 40$ (red arrows: epithelioid or glomeruloid islands, green arrows: enlarged lymphangion); (b) HE $\times 100$; (c) HE $\times 200$; (d) AAT+; (e) CD31+; (f) CD34+; and (g) Fli-1+.

whereas they showed no effect toward the patient. PLT counting decreased to $15 \times 10^9/L$. Although consumptive coagulopathy was reversed by with transfusions of PLT on the seventh day, the PLT still decreased significantly the next day. Due to the ineffectiveness of the drug therapy, intractably consumptive coagulopathy, and the worsening of breath constriction caused by the mass, we proposed to perform a surgical resection of the patient's KHE. Surgical resection was given under general anesthesia on the ninth day, and PLT transfusion was given 12h before the operation. During the surgery, the lesion was found to be fragile, prone to bleeding, infiltrating deep into the intercostal space, and extending to the whole anterior chest wall. The operation lasted for 145 min, with a hemorrhage of 150 mL, accompanied by transfusion of plasma, PLT, and erythrocytes. Surgical anesthesia and analespsia were successful. After the surgery, PLT counting was $131 \times 10^9/L$, $180 \times 10^9/L$,

and $157.6 \times 10^9/L$ during the first 3 days, respectively, and the highest PLT count was $472 \times 10^9/L$ on the eighth day after operation. The graph (Graph 1) described the change of PLT pre- and post-operation. In addition, all vital signs of the patient were good, except the operative incision in his chest wall. Because of the enormous violaceous mass extending to the chest wall from above the umbilicus to the neck as well as the aggressiveness of the KHE, some skin in the chest wall had to be excised, which made the suture tensional. Pathology results demonstrated a typical KHE. As shown in Figure 2, the predominant neoplastic component of KHE recapitulated the phenotype of blood vascular endothelium (CD31 and CD34 positive), whereas the epithelioid or glomeruloid islands located within the vascular nodules appeared to represent specialized zones in which PLT trapping and blood destruction occurred,^{1,2} suggesting KHE.



Figure 3. After a 9-month follow-up, the infant looked good but had a scar on his chest. This picture was taken on 18 July 2015.

The infant was discharged after 52 days of treatment. The blood laboratory evaluations showed that the PLT was $265 \times 10^9/L$, red blood cell (RBC): $3.1 \times 10^{12}/L$, hemoglobin (HGB): 99 g/L, hematocrit (Hct): 0.31, prothrombin time (PT): 17.9 s, high international normalized ratio (INR): 1.38, thrombin time (TT): 17.00, D-dimer: 0.2 $\mu\text{g}/\text{mL}$, and fibrinogen (FIB): 2.72 g/L. The appearance of infant looked good except a scar on his chest. After a 9-month follow-up, the infant lived a healthy life with no evidence of the recurrence of disease (Figure 3).

Conclusion

KHE with KMP is a rare disease characterized by PLT trapping within a vascular tumor leading to thrombocytopenia and intravascular coagulation. The treatment of KMP is very challenging and may require a multimodality approach, such as steroids, cytotoxic agents, interferon, vincristine, radiation, and embolization.^{3–6} However, the effectiveness of these methods is variable.

The progress of KMP is very quick and always accompanied with a high mortality. In recent medical literature, the surgical resection is not recommended as the first choice, unless it is necessary or in emergent conditions. Drolet et al.⁷ published the “Consensus-derived practice standards plan for complicated kaposiform hemangioendothelioma,” in which they 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. However, the problem is that even after non-operative treatment, residual KHE often exists. They are (more or less) prominent dormant vascular tumors, not “scars” and, clinically as well as histologically, they differ markedly from involuted hemangioma.⁸ The dormant vascular tumors also possess some proliferative activity, which is still life-threatening.⁹ Notably, the procedure of surgical resection

in this study was found to be more smooth than expected—the lesions could be easily separated and excised from the normal tissue. Thus, we speculated that for this case, better therapeutic efficacy may be obtained if the surgical resection was performed immediately after the KMP was diagnosed. Compared with other nonsurgical treatments, surgical resection may be more effective with less side effects and is usually curative with a complete reversal of coagulopathy.

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Declaration of conflicting interests

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Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

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Informed consent

Written informed consent was obtained from the patient’s parents for the anonymized information to be published in this article.

References

1. Lyons LL, North PE, Mac-Moune Lai F, et al. Kaposiform hemangioendothelioma: a study of 33 cases emphasizing its pathologic, immunophenotypic, and biologic uniqueness from juvenile hemangioma. *Am J Surg Pathol* 2004; 28: 559–568.
2. O’Rafferty C, O’Regan GM, Irvine AD, et al. Recent advances in the pathobiology and management of Kasabach-Merritt phenomenon. *Br J Haematol* 2015; 171: 38–51.
3. Wang P, Zhou W, Tao L, et al. Clinical analysis of Kasabach-Merritt syndrome in 17 neonates. *BMC Pediatr* 2014; 14: 146.
4. Ryan C, Price V, John P, et al. Kasabach-Merritt phenomenon: a single centre experience. *Eur J Haematol* 2010; 84: 97–104.
5. Malhotra Y, Yang CS, McNamara J, et al. Congenital kaposiform hemangioendothelioma with Kasabach-Merritt phenomenon successfully treated with low-dose radiation therapy. *Pediatr Dermatol* 2014; 31: 595–598.
6. Nakib G, Calcaterra V, Quaretti P, et al. Chemotherapy and surgical approach with repeated endovascular embolizations: safe interdisciplinary treatment for Kasabach-Merritt syndrome in a small baby. *Case Rep Oncol* 2014; 7: 23–28.
7. Drolet BA, Trenor CC, Brandão LR, et al. Consensus-derived practice standards plan for complicated kaposiform hemangioendothelioma. *J Pediatr* 2013; 163(1): 285–291.
8. Enjolras O, Mulliken JB, Wassef M, et al. Residual lesions after Kasabach-Merritt phenomenon in 41 patients. *J Am Acad Dermatol* 2000; 42(2 Pt 1): 225–235.
9. Chu C-Y, Hsiao C-H and Chiu H-C. Transformation between Kaposiform hemangioendothelioma and tufted angioma. *Dermatology* 2003; 206(4): 334–337.