Transcatheter Arterial Embolization in A Newborn with Cervical Kaposiform Hemangioendothelioma and Kasabach-Merritt Phenomenon.

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Abstract

We report a case of a 9-day-old newborn who underwent arterial embolization for Kaposiform hemangioendothelioma (KHE) with Kasabach-Merritt phenomenon (KMP), combined with sirolimus treatment, and the outcome was favorable. To the best of our knowledge, there are no case reports of such small infants undergoing arterial embolization to treat KHE. Our successful experience of treating KHE with KMP showed that transcatheter arterial embolization is feasible and can be used as an alternative to surgical resection, even in small infants.

Introduction

Kaposiform hemangioendothelioma (KHE) is a rare vascular tumor. When the Kasabach-Merritt phenomenon (KMP) is complicated, it can be serious and requires aggressive treatment. Surgical resection is considered to be the standard method for the treatment of vascular tumors, but it may be difficult to perform due to acute bleeding and severe coagulopathy in patients complicated with KMP. Medical treatments with corticosteroids, vincristine, and sirolimus have been suggested for the management of KHE; however, drugs take too long to correct coagulopathy with active bleeding. Transcatheter arterial embolization may be an alternative therapy for KHE with KMP.

Results

The patient was a male newborn from the first pregnancy of a clinically healthy woman. He was born at 38+6 weeks of gestational age by normal spontaneous delivery, weighing 2890 gm. His mother received regular prenatal examinations revealing no specific abnormality. A large (approximately 6 x 5 x 2 cm), firm, immovable mass with purpuric and bruised appearance was found over his left postauricular site (**Figure 1A**). He was initially diagnosed with infantile hemangioma and treated with propranolol. However, although gauze soaked with epinephrine was used locally and directly pressed to stop the bleeding, hemorrhage from the hemangioma continued from the sixth day of birth. Laboratory data showed thrombocytopenia (platelet of 17000/uL), coagulopathy (international normalized ratio of prothrombin time was 2.04, activated partial thromboplastin time was greater than 150 sec) and hypofibrinogenemia (fibrinogen of 152.1mg/dL). These findings are consistent with Kasabach-Merritt phenomenon (KMP). Magnetic resonance imaging revealed a vascular-rich lesion with central scars, consistent with Kaposiform hemangioendothelioma (KHE) (**Figure 2A**). Despite sirolimus 0.8 mg/m^2 twice daily and transfusion with fresh frozen plasma, cryoprecipitate, platelets, the bleeding could not be stopped. Because of KMP with active bleeding and surgical operation

not being feasible, transcatheter arterial embolization was performed at 9 days of age and 3.05 kg. Selective left common carotid arteriography demonstrated a single feeding artery to the mass lesion without collateral artery to adjacent organs (**Figure 2B**). A 4F delivery catheter (Amplatzer Judkins Right catheter, Abbott Medical, MN, U.S.A.) was advanced over a 0.018" Terumo guide wire into the feeding artery of the mass lesion. A total of three Cook Embolization Coils, two 3 mm x 4 cm, one 3 mm x 5 cm, and one 3/2 mm x 2 cm Cook Tornado Embolization Microcoil (Cook Medical, Bloomington, IN, USA), were selected for embolization of the feeding artery. Repeat arteriography showed no obvious residual shunt after embolization (**Figure 2C**). The bleeding was stopped, and he no longer needed a blood transfusion on the second day after embolization. Follow-up laboratory data revealed neither thrombocytopenia nor coagulopathy 1 week later. He was discharged from hospital on day 18 (**Figure 1B**). At 3 months, the KHE had almost completely regressed (**Figure 1C**), and then sirolimus was discontinued.

Discussion

KHE is a rare vascular neoplasm during infancy and should be carefully differentiated from infantile hemangioma[1]. The prevalence of KHE in Massachusetts is about 1 in 100,000 children[2]. It can present as a raised subcutaneous mass, involving the extremities, trunk, or cervicofacial region. In addition, it may present as intrathoracic or retroperitoneal lesions in 10% of patients, not involving the skin[2]. The major pathological manifestations of KHE are abnormal angiogenesis and lymphangiogenesis[3]. Diagnosis is based on the combination of clinical, histologic, and imaging features. MRI can illustrate an ill-defined mass, hypointense or isointense compared with muscle in T1-weighted sequences, and hyperintense in T2-weighted sequences[4]. Although KHE is a benign vascular tumor that does not spread to other parts of the body, it can grow aggressively and is often associated with the most serious complication, KMP, which is a life-threatening complication of KHE. The KMP was first described in 1940 with characteristics of active consumptive coagulopathy[5]. Therefore, laboratory data can show profound thrombocytopenia and hypofibrinogenemia with elevated markers of coagulation activation. In a large cohort study[2], the risk factors for KMP included the tumor location (intrathoracic or retroperitoneal involvement), the tumor depth (infiltration into muscle or fascia), and the tumor size (maximum diameter greater than 8 cm). Immediate management is needed to avoid high mortality and morbidity. It is currently thought that KMP occur in approximately 70% of KHE, but not in infantile hemangioma[4,6]. Given the high mortality range from 8 to 24%[4,7], accurate diagnosis and prompt treatment are important. However, there is no standard treatment guideline due to the rarity of KHE.

The treatment method for KHE is mainly based on the opinions of experts, and a small number of case reports and case series. Supportive treatment should be provided first to prevent the patient's condition from getting worse, followed by curative therapy for the underlying vascular tumor. Fresh frozen plasma or cryoprecipitate is used to correct coagulopathy. Platelets are reserved for active bleeding or before surgery, because it will exacerbate mass bleeding and platelet trapping in the tumor[3,8]. Although surgical resection is considered to be the standard method for the treatment of vascular tumors, it is difficult to perform during the active phase of KMP due to acute bleeding and severe coagulopathy. Since most KHE is infiltrative and may invade adjacent organs, it is almost impossible to obtain a clear tumor margin for surgery[9]. Arterial embolization may be an effective treatment method for KHE combined with KMP, but its main limitation is the technical difficulty of accessing the very small feeding arteries in neonates and young infants[3]. Previous studies have found that arterial embolization only had a temporary therapeutic effect for some cases of KMP[7]. According to the 2013 consensus of medical treatment for complicated KHE[9], intravenous vincristine 0.05 mg/kg once a week and oral prednisolone 2 mg/kg/day or intravenous methylprednisolone 1.6 mg/kg/day are recommended as the first-line treatment for KHE patients with KMP, and oral prednisolone 2 mg/kg/day is recommended for those without KMP. Sirolimus has recently been suggested as an alternative therapy for KHE [3,8]. Cashell J, et al. reported the life-saving treatment of an infant with KHE and KMP after three weeks of treatment with prednisolone, vincristine, and sirolimus[10].

In the acute stage of KMP, it is difficult to perform surgery when there is evidence of bleeding tendency. On the other hand, drugs take too long to correct coagulopathy with active bleeding. Transcatheter arterial

embolization may be an alternative therapy. However, to the best of our knowledge, there are no case reports of such small infants receiving arterial embolization to treat KHE. In the present case, a 9-day-old newborn underwent arterial embolization for KHE with KMP, combined with sirolimus treatment, and the outcome was favorable. Our successful experience of treating KHE with KMP revealed that transcatheter arterial embolization is feasible and can be used as an alternative to surgical resection, even in small babies.

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Conflict of Interest

The authors declare that there is no conflict of interest.

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Legends of Figures

Figure 1. A, A large (approximately 6 x 5 x 2 cm), hard, immovable mass behind the left ear with purpuric and bruised appearance. B, The mass shrank after 2 weeks of arterial embolization. C. The mass was almost completely regressed at 3 months of age.

Figure 2. A, Magnetic resonance imaging showed a vascular-rich lesion with central scars. B, Selected left common arteriography demonstrated a single feeding artery to the mass lesion without collateral artery to adjacent organs (arrow). C, Repeat arteriography revealed no residual shunt after transarterial embolization using multiple coils (arrow).