**Case Report**

**Upper arm life-saving amputation of a 12 day-old neonate due to extensive vascular tumor of the upper extremity**

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**ABSTRACT**

Vascular tumors in neonates are mostly benign; however, locally aggressive voluminous forms may destabilize the hemodynamics of a neonate. Herein, we present an unusual case of a neonatal giant vascular tumor in the right upper extremity, causing a consumption coagulopathy and acute deterioration of vital signs. The patient required mechanical ventilation, inotropic support, and administration of blood products by the seventh day. Vascular embolization attempts failed to improve the general condition of the patient. Due to the deteriorating and life-threatening general condition of the patient, amputation around the upper arm level occurred under emergency conditions on the twelfth day. The patient’s hemodynamic parameters were regained immediately, with neither inotropic agents nor blood products required after the second postoperative day. Clinical and pathological diagnosis revealed kaposiform hemangioendothelioma. Patient monitoring proceeded until the age of 15 months, with no local recurrence around the stump or soft tissue coverage complications. Therefore, since other treatment options failed, the early amputation decision was life-saving.

**Case Presentation**

A neonatal female patient was examined by the orthopaedic surgery team in the neonatal intensive care unit on the first postnatal day due to her abnormal right upper extremity (Figure 1).

The patient was delivered by a 25 years old mother through caesarean section in the 35th week of gestation. Review of the prenatal history revealed that the upper extremity abnormality was detected by Obstetricians in the 33rd week of gestation, during a routine fetal ultrasonography control. Magnetic resonance imaging (MRI) results indicated the presence of a vascular tumor or malformation; malformations and tumors. Among these, the locally aggressive tumor, kaposiform hemangioendothelioma (KHE), can cause a severe and life-threatening consumptive coagulopathy termed the Kasabach-Merritt phenomenon (KMP) (1-3). In this study, we present and discuss a unique neonatal case of KHE affecting the entire right upper extremity, which required life-saving amputation on the twelfth postnatal day.

Vascular anomalies are classified into two main groups; malformations and tumors. Among these, the locally aggressive tumor, kaposiform hemangioendothelioma (KHE), can cause a severe and life-threatening consumptive coagulopathy termed the Kasabach-Merritt phenomenon (KMP) (1-3). In this study, we present and discuss a unique neonatal case of KHE affecting the entire right upper extremity, which required life-saving amputation on the twelfth postnatal day.

Although physical examination suggested normal vital signs, her right upper extremity was about 4-fold wider when compared the left side, with the extremity exhibiting a cyanotic-like colour. The enlargement and abnormal colour extended from the middle of the clavicle to the fingers (Figure 1), with no active motion observed at the abnormal extremity. Through auscultation, noise was audible on the medial side of the upper arm. Clinical and gross features examination of the mass produced a preliminary diagnosis of a benign aggressive vascular tumour, considering the classification of the International Society for the Study of Vascular Anomalies (ISSVA) (1-3). The patient exhibited no hemodynamic problems, but thrombocytopenia was detected from her complete blood counts in the early postnatal period. A Doppler ultrasound and MRI of the patient’s arm revealed the entire upper extremity, from the shoulder to the fingers, was affected by a vascular tumor or malformation. On the second postnatal day, serious throm-
bocytopenia (9000/mm$^3$), prolongation of the coagulation time, and high D-dimer levels indicated the development of a consumptive coagulopathy. Fresh frozen thrombocyte and thrombocyte suspensions were administered, and at this time, no clinical or echocardiographic sign of heart failure was noticed. The interventional radiology team attempted vascular embolization on the fourth postnatal day, but this failed to halt progressive deterioration of the patient’s general condition. On the seventh postnatal day, development of lethargy, hypotonia, and hypotension accompanied by deep metabolic and respiratory acidosis made intubation of the patient necessary. High frequency oscillatory ventilation (HFOV) was initiated after conventional ventilation techniques failed to increase oxygen saturation. At that instant, laboratory tests relieved the sustained thrombocytopenia, abnormal coagulation parameters, and anaemia (haematocrit was 25), without abnormality in the infection parameters. The administration of fresh frozen plasma including thrombocyte and erythrocyte suspensions and fluid support failed to alleviate the hypotension, triggering increasing use of dopamine, dobutamine, and adrenaline. A second attempt for vascular embolization occurred on the tenth postnatal day, with no beneficial effect on the patient’s general condition. On the twelfth postnatal day, severe deterioration of the patient’s general condition was evident through loss of consciousness, weakened pulses, prolonged capillary refilling time (>4 seconds), and decreased urine output (1 mL/kg/hour). The mean airway pressure was increased to 20 cmH$_2$O on HFOV to normalize patient’s oxygen saturation with an oxygenation index >30. Laboratory tests revealed increased serum liver enzymes, direct bilirubin, and high urea and creatinine levels in addition to abnormal coagulation tests. These results indicated emerging multi-organ failure, and so, an emergency upper arm amputation was planned, hoping to save the patient’s life. On the amputation day, the entire body was cyanotic, with necrosis plaques on the palmar side of the hand and over the dorsal surface of the distal forearm (Figure 3). High upper arm amputation was performed under general anaesthesia. Since the skin was affected by the vascular mass, a monopolar electro-cautery pen served in cutting the skin incisions. Amputation was completed by clamping, ligation and cutting of the abnormal deep structures successively. The patient was extubated on the second postoperative day characterized by quick recovery of the patient’s clinical condition. No inotropic or other medication support was needed by the patient after the amputation and no wound complications or infection developed at the stump. The last postoperative examination showed decrease in the preoperative protuberance and discolouration of the shoulder region (Figure 4).
Histopathological and immunohistochemical evaluations were conducted on samples from different levels of the amputated upper extremity. The dermis, subcutaneous tissue, muscles, and bones were extensively affected by a lobular capillary proliferation. Although less than the lobular type, tissues were also affected by a diffuse type capillary proliferation. In sections from the subcutaneous tissue, areas of slit-like lumens of proliferative capillaries lined by spindled cells and dilated vessels (Hematein-eosin stain; X100) existed (Figure 5) (2). Vascular endothelial cells showed positive staining with CD34, CD31, and FLI1 in the histoimmunochemical study, while staining with D2-40 and GLUT-1 was negative. These clinical and pathological data provided the final diagnosis as KHE complicated by Kasabach–Merritt Phenomenon (KMP) (1-3).

Discussion

Kaposiform hemangioendothelioma is a rare benign aggressive vascular tumor. It can be fatal in infants due to its profound coagulopathy known as KMP. Although other types of vascular tumors like tufted angioma and rapidly involuting congenital haemangio- ma (RICH) are also associated with coagulation disorders, no other tumor type is reported to be associated with severe KMP as KHE (1, 3). Therefore, the presence of KMP enhances the clinical diagnosis of KHE for vascular tumors. Definitive diagnosis is achieved by histopathologic findings from biopsy or surgical resection (2, 3), although differential diagnosis of vascular anomalies solely based on histopathology studies are difficult due to similarity of the findings (2). The treatment of complicated KHE is difficult, but surgical resection or amputation is often not preferred in the early stages of clinical deterioration, since high surgical risks exists for infants with unstable hemodynamics. Some of these risks are worsening of the KMP due to manipulation of the tumor, local or systemic hemorrhage, mortality, and impossibility of total resection in tumors with no clear surgical margins (3). In the case studied, in addition to medications, embolization of the abnormal vessels was attempted twice before the surgery. Despite these efforts, worsening of the clinical condition necessitated emergency surgical intervention. We thought that removal of the life-threatening lesion could only be possible by amputation of the affected upper extremity. In the literature, although a few cases of life-saving extremity amputations or vascular ligations are reported (3, 4), no case of amputation as early as in our study is available. The studied case was definitively diagnosed as KHE, based on clinical and pathological findings.

In the studied case, striking highlights of the surgery are the critical health condition of the 12 days-old infant and the massive hemorrhage from all tissues of the amputated site. Due to these features, almost all skin incisions were performed using electro-cautery, with deep tissues clamped and ligated massively before cutting. Although clinical, radiological, and intra-operative data reveal that the lesion extended proximally anterior to the shoulder joint, the proximal humerus was chosen as the amputation site. We decided that a more proximal and complicated surgical intervention could be mortal for the patient, who was in critical condition. The relatively less affected skin of the posterior side of the shoulder served in covering the stump, and the wound healed without any problem after surgery.

Conclusion

Massive attacks of the right extremity by a vascular tumor disrupted the hemodynamics in a neonate studied. If medical treatment fails, timely surgery should be considered as a life-saving intervention. Although risky, surgical intervention at an appropriate time and manner may prevent fatal progress and improve the clinical condition. In our study, the clinical condition of the infant deteriorated rapidly in the first postnatal week due to a massive upper extremity vascular tumor and its associated coagulopathy. This case required a very early upper arm amputation.
Informed Consent: Written informed consent was obtained from patient who participated in this study.

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