

A newborn with Kasabach–Merritt syndrome

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Case report

A term baby with a birth weight of 2.5 kg was born to a primigravida mother. There were no obvious abnormalities on clinical examination of the baby except for a reddish brown or blue, tender skin lesion that rapidly evolved into a violaceous, bulging mass roughly measuring 6 cm x 7 cm on the antero-lateral aspect of the right thigh with petechiae and purpura locally (Figure 1).



Figure 1: Large vascular tumour of right thigh

The swelling was fluctuant. There was no purpura or petechiae in other areas of the body and no history of a previous vascular lesion. The baby had a normal leucocyte count but had thrombocytopenia (75,000/cu mm). Baby's activated partial thromboplastin time was 76.2 seconds (normal 42.9 ± 5.80 seconds), prothrombin time 29.4 seconds (normal 13 ± 1.43 seconds) and international normalized ratio 3.2 (normal range 1-1.5). Fibrinogen level was 18.4 mg/dl (normal values

200–400 mg/dl), fibrin degradation products level was 148.6 mg/L (normal <10 mg/L) and d-dimer level was 17.3 mg/L (normal value less than 0.5 mg/L). Skin biopsy of the lesion showed dilated irregular thin walled vascular channels lined by flattened endothelial cells in the dermis. Colour Doppler showed feeding arterial supply to the swelling. Baby was diagnosed as Kasabach–Merritt syndrome (KMS). Coagulopathy was treated with fresh frozen plasma and platelet transfusion. The patient was started on prednisolone 3mg/kg/day for 4 weeks. However, there was no regression in the tumour size and thrombocytopenia persisted. Embolization of feeding vessel was not practically feasible at our centre. Propranolol was tried but patient developed bronchospasm and hence this was discontinued. Other treatment options considered were vincristine and interferon-alpha-2b, but these could not be administered due to financial constraints of patients' family. Subsequently, patient was referred to a higher tertiary level centre, which had vascular surgery/interventional radiology expertise.

Discussion

KMS is an uncommon but clinically important entity since the mortality attributed to it is roughly 30%^{1,2}. To the best of our knowledge, only around 200 cases of KMS have been reported so far^{1,2}. This is a consumptive coagulopathy which leads to thrombocytopenia and also defects in clotting factors leading to disseminated intravascular coagulation and even death³. It is usually associated with kaposiform haemangioendothelioma, tufted angiomas and rarely congenital hemangiomas⁴. Other clinical features include abdominal distension because of haemangiomas in liver causing hepatomegaly, jaundice, petechiae, bruising, bleeding manifestation and anaemia. Patients uniformly show severe thrombocytopenia, low fibrinogen levels, high fibrin degradation products (due to fibrinolysis), and microangiopathic haemolysis⁵. Different interventions are recommended including the use of steroids, dipyridamole, pentoxifylline, ticlopidine, and heparin, vincristine, interferon (IFN), compression, embolization, laser therapy, sclerotherapy, radiotherapy, and embolization of the feeding vessel of the tumour^{6,7}.

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