

Images in Clinical Practice

Kasabach-Merritt Syndrome

A male baby weighing 3620 g was born to a primigravida mother at 38 weeks gestation by Caesarean section. Antenatal USG had revealed a large hypoechoic mass over the left trunk and lower limb with poor visualization of the left lower limb. At birth, the baby had marked pallor, poor perfusion and a large vascular tumor involving the entire left lower limb with a few ecchymotic patches on the lower abdomen (*Fig. 1*). This was probably a hemangioma or a giant arteriovenous malformation. Investigations revealed hemo-

globin 11.4 g/dL, platelet count 63,000/cu mm. Prothrombin time 43 seconds and APTT 53.1 sec, and d-Dimer was positive (500 mcg/L). A diagnosis of Kasabach-Merritt syndrome with disseminated intravascular coagulation was made. The infant required ventilatory support, plasma and packed red cell transfusions and inotropic support. The large size and situation of the tumor precluded surgical management. While other interventions were being considered, baby developed refractory circulatory failure and died on sixth day of life.

Kasabach-Merritt phenomenon is seen in



Fig. 1.

children with large vascular tumors and is characterized by thrombocytopenia, consumption coagulopathy and microangiopathic hemolytic anemia. The clinical presentation can be severe anemia, torrential hemorrhage, rapid increase in size of the tumor or high output cardiac failure due to the arteriovenous malformation, with an overall mortality of 20-30%. The vascular tumor is believed to be a tufted angioma or a kaposiform hemangio-endothelioma and not a true hemangioma. The vascular lesion can be superficial or visceral occurring in thoracic, abdominal, pelvic or intracranial sites. Thrombocytopenia persists for a variable period of time lasting from a few months to years. The child may die of infection or hemorrhage.

Treatment is supportive and includes administration of platelet, red cell and plasma transfusions. Digitalization may be required for high output cardiac failure. Various drugs such as systemic steroids, recombinant interferon alpha, vincristine and cyclophosphamide have been used with variable success. Other modes of management include surgical excision, arterial embolisation and radiotherapy.

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Systematized Nevus Depigmentosus

A 2-year-old girl, product of non-consanguineous marriage, presented with hypopigmented patches over the entire body since 5 months of age. New lesions appeared over the next eight months, after which they have been static. There was no history of visual or hearing deficit, seizures, development delay, abnormal sweating or inflammatory changes preceding pigment loss. She had normal scalp hair and teeth. Family history was not contributory. Dermatological examination revealed multiple, variable sized, oval to irregular, bilaterally distributed hypopigmented macules, scattered over the

back, chest, abdomen, upper and lower limbs and sparing the scalp and face (*Fig. 1*). There was no leucotrichia. Systemic examination including musculoskeletal, ophthalmological and neurological was normal. Radiological examination including chest X-ray, ultrasound of the abdomen and pelvis, X-ray skull, pelvis, hands and feet were normal. However, X-ray of the cervical and thoracolumbar spine revealed spina bifida of the C5, C6 and C8 vertebrae. A diagnosis of systematized nevus depigmentosus (ND) with spina bifida was made.

ND is a congenital, non familial, well circumscribed, uniformly hypopigmented macule, stable in its relative size and distribution throughout life and involving predominantly the trunk and proximal