

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

extremities and rarely the head and neck. Both sexes are equally affected and there is no distinct pattern of inheritance. Histologically there are normal to decreased number of melanocytes with normal size, shape and melanin content of melanosomes, but reduced number and aggregated within vacuoles. Clinical diagnostic criteria for ND include (i) leukoderma present at birth or onset early in life, (ii) no alteration in distribution of leukoderma throughout life (iii) no alteration in texture, or change of sensation, in the affected area and (iv) no hyperpigmented border around the achromic area. Three morphological variants of ND have been described; isolated (circular or rectangular), which is the commonest presentation, segmental and the rare systematized (unilateral whorls or streaks) variety. Systemic manifestations are rare in ND, although systematized ND in association with seizures, mental retardation, limb hemihypertrophy, atopic dermatitis, yellow scalp hair, has been described. Systematized ND is often confused with hypomelanosis of Ito (incontinentia pigmenti achromians). It presents as unilateral or bilateral hypopigmented streaks and whorls that follow Blaschko's lines, the lesions exhibit changes in their manifestation over time (initially progress and then regress), are associated with ocular, musculoskeletal and CNS abnormalities in 62% to 94% of cases and show familial tendency (autosomal dominant mode of inheritance in some cases). Other differential diagnosis in children who present with hypopigmented macules include vitiligo, piebaldism, nevus anemicus, ash-leaf macules



Fig. 1. Systematized nevus depigmentosus.

of tuberous sclerosis, pityriasis alba, pityriasis versicolor, hypopigmentation of post kala-azar dermal leishmaniasis (PKDL) and post inflammatory hypopigmentation.

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