Hallerman-Streiff Syndrome

A two-month-old female child presented with congenital teeth and sparse of hairs over face and scalp since birth. She was a product of nonconsanguinous marriage, full term delivery, weighing 2.75 kg at birth.

On examination, upper right canine and left upper central incisor were erupted which appeared normal. The tongue was slightly big with high arched palate. Hairs over the scalp, eyebrows, and lower eyelids were almost absent but normal on both upper eyelids. Skin over the central portion of the face was slightly atrophic, shiny, and revealed multiple telangiectasias (Fig. 1) mainly over the bilateral supraciliary arches. Mild micrognathia was also noted. Ophthalmologic examination revealed bilateral congenital cataract and microphthalmia. A diagnosis of Hallerman-Streiff syndrome (HSS) was made.

The majority of the cases of HSS are sporadic. Autosomal dominant inheritance has been suggested but the current data indicates autosomal recessive pattern. The classical features of HSS include short head, hypoplastic mandible and beaked nose giving parrot-like appearance. A characteristic pattern of alopecia along the lines of cranial sutures has been described. Atrophy of the facial skin with telangiectasia, congenital cataract, and congenital teeth are other characteristic features of this syndrome.

At times, it needs to be differentiated from progeria. The presence of large cranium with prominent scalp and thigh veins, sclerodermoid changes on lower trunk and thighs, senile looks, and cardiovascular and musculoskeletal involvement can differentiate progeria from HSS.

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Fig. 1. Sparsening of hair, atrophy with telangiectasia and microphthalmia.