CASE REPORT

Accelerated phase of Chediak-Higashi syndrome-a case report with review of literature

Hephzibah Rani, Deepak Kanabur

SDM COLLEGE OF MEDICAL SCIENCES & HOSPITAL, DHARWAD,INDIA

Abstract:

The Chediak Higashi Syndrome (CHS) is a rare autosomal recessive disease characterized by partial oculo-cutaneous albinism, frequent pyogenic infections, presence of giant granules in leucocytes and other granule containing cells. Associated findings include silvery hair, photophobia, horizontal and rotatory nystagmus, hepatosplenomegaly and peripheral neuropathy. Mutation of the LYST gene defines the syndrome. The first case of Chediak Higashi Syndrome was reported in 1943. Since its first description, around 170 cases have been reported in the literature till date and 10 cases have been reported from India.

Keywords *Chediak-Higashi, accelerated phase, abnormal granules*

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Corresponding author*:* *Hepzibah Rani, Assistant Professor in Pathology, SDM College of Medical Sciences-Hospital, Dharwad, India. dr.hephzibah@gmail.com*