Vogt Koyanagi Harada Syndrome

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Vogt Koyanagi Harada Syndrome (VKHS) is an uncommon systemic syndrome characterized by intraocular inflammation with associated cutaneous, auditory and neurological abnormalities(l-4). This condition is common in the third and fourth decade, rarely affecting young children. We report this rare syndrome in an 11 year old female.

Case Report

An 11-year-old female girl presented to us with fever of 3 months duration. Additionally she had vitiligo for two years and deafness and alopecia for the last 3 months. Two years ago, the patient had an episode of pain, watering and congestion of both eyes (suggestive of nontraumatic uveitis) for 2 weeks. She responded to therapy in the form of eye drops and oral medication (Ayurvedic medicine). A few months later she developed patches of vitiligo over her face, chest, and legs and white hair of scalp and eye lashes (poliosis) *(Figs. 1 & 2)*.

On examination, she had vitiligo patches, poliosis, deafness and the rest of the systems including the eye were essentially normal. The patient was thoroughly investigated for fever. Complete blood counts, *Mantoux test,* X-rays chest and abdomen, USG abdomen and ANA/DsDNA were normal. On brain stem evoked response audiometry bilateral sensorioneural deafness was confirmed and skin biopsy from hypopigmented patches revealed the absence of melanocytes.

The patient was appropriately treated for urinary infection. Two weeks after admission she had meningoencephalitis with pleocytosis on CSF examination. In our patient, in view of nontraumatic uveitis (which possibly responded to steroid containing preparation), vitiligo, poliosis, deafness and aseptic meningitis, the diagnosis of Vogt Koyanagi Harada syndrome was entertained although the presentation was atypical as aseptic meningitis had occurred rather late in the course of the illness, after the ophthalmic phase.

Later, unfortunately the patient had a very stormy course of events. She developed peritonitis with perforation. Exploratory laparotomy revealed multiple ulcers in the distal ileum which on histopathology confirmed the diagnosis of tuberculosis. She was given anti-tubercular treatment without steroids to which the response was good. Eight months after follow-up, the patient has vitiligo and poliosis but no ocular findings or sequelae and hence steroid therapy was not indicated at this stage.

Discussion

Vogt Koyanagi Harada Syndrome (VKHS) is a rare autoimmune multisystem syndrome. It was originally described as separate entities: namely anterior uveitis, vitiligo, poliosis and deafness comprised the Vogt component, whereas posterior uveitis consisted the Harada disease. Pres-

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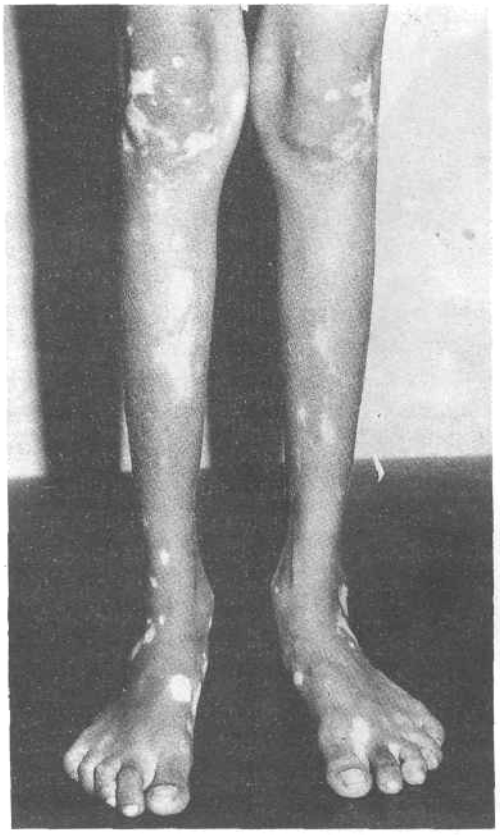

Fig. 1. Depicts bilateral symmetrical vitiligo patches over the lower limbs.

ently all the above manifestations are considered as a continuum of the same illness and are referred to as VKHS(1,2,5). The peak age of presentation is the third and fourth decade with equal sex incidence. Cases have most often been reported from Japan, North and South America and Europe with a seasonal peak during Spring and Autumn(l,4).

This syndrome is presumably viral in nature with autoimmune phenomenon that results in destruction of melanocytes especially of the uveal tract, meninges, cochlea, andepidermis(l,4-6). VKHS occurs in three phases namely the prodromal phase or the meningoencephalitic phase, followed a week or two later by the ophthalmic phase of bilateral uveitis (anterior and/or posterior) with auditory impairment occurrings in 50%. The spectrum of other ocular changes includes iridocyclitis, vitreitis and papillitis(3). During the last or the recovery phase, 80-90% develop poliosis (of brows, eyelashes and scalp), 60-70% develop vitiligo (usually symmetrical) and 50-75% have alopecia areata or diffuse hair loss(l,2,4).

The association of vitiligo, poliosis and ocular changes clearly differentiates this syndrome from any other. Alezzandrini's syndrome is a close differential which is commonly seen in adolescents and is characterized by unilateral impairment of vision and ipsilateral facial vitiligo(4,5). The pigmentary changes in VKHS tend to be permanent though some authors have documented clearance after chlorambucil therapy(3). Hearing is usually completely restored and uveitis may take a year or more to recover and in 66% visual acuity returns to normal(4).

Therapy in VKHS is targeted towards management of uveitis to avoid complications like glaucoma, cataract and subretinal neovascular membranes(2,3). Early institution of steroids and in resistant cases, combination with chlorambucil and cyclophosphamide is advisable. Hence early diagnosis of ocular changes is crucial and can be enahanced by using Indocyanine green angiography, Ultrasound microscopy and MRI(2,7).

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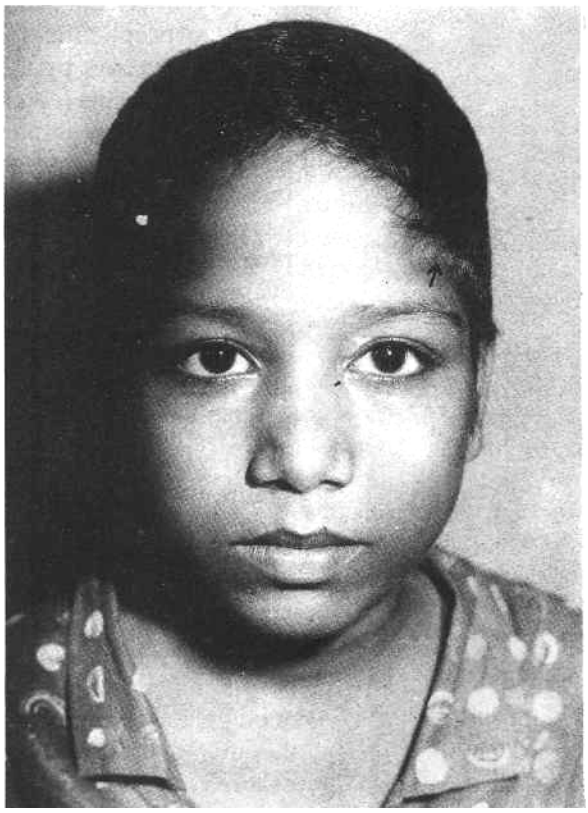

Fig. 2. This figure eveals the poliosis of scalp hair (marked with arrow) and a vitiligo patch on the left eyelid

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