Case Reports

Vogt-Koyanagi-Harada syndrome

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The Vogt-Koyanagi-Harada (VKH) syndrome is a rare, chronic bilateral, granulomatous panuveitis with extra ocular, manifestations in the central nervous system, auditory system and intergumentary system. Ophthalmologists rather than dermatologists report most of the patients with VKH syndrome. Alopecia, poliosis and vitiligo are common manifestations of this syndrome. We report a case of 75-year old man with VKH syndrome, who developed bilateral deafness, tinnitus, alopecia, poliosis and vitiligo following an episode of bilateral panuveitis.

Case Report

A previously well, 75 year old man presented to the ophthalmologist with sudden onset blindness and severe headache. The headache gradully subsided in 2 weeks. His blindness responded to topical steroids and homatropine eye drops. Three months later his vision was further impaired. Two weeks later he developed hearing loss, tinnitus, diffuse hair loss, graying of eyebrows and 'white spots' over his forehead (Fig. 1), dorsum of his hands and legs. Examination of his scalp revealed a diffuse, patchy, non-scarring alopecia. The patient was noted to have graying of eyebrows, hypopigmented macules on either side of his forehead, dorsum of his hands and leg. Examination of his ears revealed bilateral sensoneural hearing loss and tinnitus.

The histology of hypopigmented macules revealed less number of melonocytes by immune stain for S 100 suggestive of vitiligo.

The diagnosis of Vogt-Koyanagi-Harada syndrome was made clinically based on the manifestions of uveitis, sensoneural hearing loss, tinnitus, alopecia, poliosis and vitiligo.

Subsequent to the diagnosis, his eye symptoms worsened. He responded to oral prednisolone 10mg daily with a reducing dose regime over several months. In addition, his hearing improved, tinnitus disappeared and alopecia improved with marked hair regrowth.

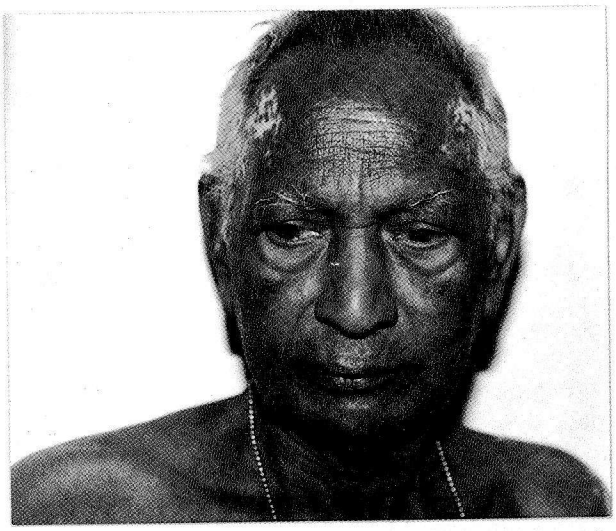

Figure 1. Vitiligo patches on the forehead and poliosis of eyebrows

Discussion

The VHK syndrome is a systemic inflammatory condition, consisting of uveitis with non-ocular manifestations affecting the central nervous system, inner ear and skin. It was first described by Vogt in 1906[1]. Koyanagi subsequently described a group of patients with Vogt-Koyanagi syndrome who had bilateral anterior uveitis, dysacousia, vitiligo, alopecia and poliosis[2]. In 1926 a similar syndrome termed Harada'a disease was described in Japan. These patients had bilateral posterior uveitis, exudative retinal detachment and cerebrospinal fluid pleocytosis[3]. In 1939, the term Vogi-Koyanagi-Harada syndrome was coined, to encompass all the features of this condition. Mean age of disease onset was 34 + 14 years. (range 8 to 75 years)[4]. There is a statistically significant

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difference in the mean age at presentation between the various ethnic groups, with Asians presenting at a significantly older age than all other ethnic groups. It occurs equally in both sexes. There may be genetic predispositions. Although, the disease has a worldwide distribution, it is more common in Asians, especially the Japanese. The exact etiology of the VKH syndrome is unknown. It is considered an autoimmune disease in which there is an attack against the melanocytes in the skin and other body tissues such as the uvea, meninges and inner ear. Classically, patients undergo three phases of clinical progression. The initial phase consists of a prodrome of fever malaise, headache, nausea, and vomiting reflecting encephalitic or meningeal involvement. Our patient had severe headache at the onset of his illness. One to two weeks after the prodrome phase, the ophthalmoauditory phase occurs consisting of bilateral uveitis, choroiditis and optic neuritis. Our patient had bilateral panuveitis with clinical manifestation of loss of vision. As the ocular manifestations subside, more than 75% of affected patients experience dysacousia, tinnitus and hearing loss. Our patient developed tinnitus and impaired hearing with the onset of visual symptoms. As the uveitis begins to subside (usually within the first 3 months), skin manifestations usually occur. Our patient developed alopecia and vitiligo subsequent to the uveitis. Appoximately 90% of affected patients develop poliosis, which may limit to the eyebrows and lashes or may involve scalp and body hair more extensively. Vitiligo develops in 63%. according to one study. Alopecia is also common, affecting 73% of patients. The exact nature of the alopecia is not well understood. In contrast there is a significant amount of information about eye manifestations in the VKH syndrome. Perhaps this discrepancy is due to the fact that most cases present and are reported by the ophthalmologists, rather than the dermatologists. Our patient experienced a sudden onset of diffuse hair loss associated with white hair replacement. On examination he had features of alopecia areata. There was a diffuse non-scarring alopecia with 'exclamation mark' hairs and vallus hair regrowth. Furthermore our patient's alopecia improved significantly with the introduction of systemic steroids.

In conclusion our patient had features of alopecia areata, vitiligo and poliosis in the setting of VKH syndrome. Rarity of this condition prompted us to report this condition.

References

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