Vogt-Koyanagi-Harada Syndrome in Siblings
(with a Brief Review of the Literature)

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Abstract. We report a brother and a sister with Vogt-Koyanagi-Harada syndrome. The girl had all the manifestations of this condition; her brother was less severely affected. Both showed mental and growth retardation. The literature on this syndrome is reviewed.

Key words: Vogt-Koyanagi-Harada syndrome – Uveitis – Vitiligo.

Introduction

The Vogt-Koyanagi-Harada syndrome (VKH) is a rare systemic disorder of uveitis, dysacusia, vitiligo, premature graying of the hair, eyebrows and eyelashes, and meningoencephalitis [9]. The first case was described by Vogt in 1906. Koyanagi in 1929 reported similar cases, after which the entity was identified as Vogt-Koyanagi syndrome [9]. In 1926 Harada reported a patient with an idiopathic uveitis affecting the posterior segment with retinal detachment and meningeal irritation [3, 8]. This condition, known as Harada disease, was predominantly seen in adults. At present, these two disorders are considered variations in severity of a single entity [9]. The similarities between this syndrome and sympathetic ophthalmia have been noted [12].

The cause of VKH is unknown [3, 5, 7, 9, 10, 13]. It is suspected that this is a virus disease or an autoimmune reaction to uveal pigment [13]. Three phases of the condition are recognized: meningoencephalitic, acute ophthalmic, and convalescent [2, 4].

This syndrome is seen mostly in 19-46-year-old adults, rarely in children. Almost all are sporadic cases; familial cases are rare. In 1932 Salus [11] reported the condition in two sisters. In 1951, Benedict and Benedict [1] described uveitis, poliosis and alopecia in male and female sibs. We present two new familial cases suggesting that the condition is inherited, probably as an autosomal recessive.

Reports of Patients

Patient 1. AA, a 13-year-old girl was seen for loss of vision—her mother was unable to give a specific age of onset. There was no history of injury or known infection of either eye. When she was in primary school, her mother noticed that she brought books very close to her eyes when reading and could see no further than 30-40 m. One week before coming to the hospital, she had a viral (?) infection and for 3-4 days her vision was reduced to light perception only. From the onset of her visual problems, her family also noticed that the patient had patches of white hair on her scalp. The eyebrows and eyelashes became thin and lighter in color and some white cilia were observed on her lids. At 6 or 7 years some white spots appeared on her trunk and limbs and became progressively larger. For the preceding 2 years deterioration in her hearing had been noted. She has not learned to read or write.

The patient’s mother (35 years) and father (42 years) are first cousins, and except for her brother no other relative is affected (Fig. 1).

On examination at 13 years her weight was 27 kg (N 50.1 ± 8.5), height 137 cm (N 159.5 ± 6.3), head circumference 53 cm (N 53.0 ± 1.4). She could hear only when spoken to very loudly. There were patches of white hair on the scalp in both occipito-parietal areas. Her eyelashes and eyebrows were thin and lighter than the scalp hair and there were some white cilia on the upper lids.

The pupils were isochoric and light reflexes were normal. The vision could not be measured as the child was uncooperative. However, it seemed that the vision in each eye was limited to light perception with just enough vision to differentiate persons. There was an abducens palsy on the left. On the right vacuolization was observed on the posterior part of the lens, and there was retinal detachment on the nasal side of the fundus on the left eye. Since there were numerous cells in the vitreous, the fundus could not be...
Patient 2. AA, the 11-year-old brother of patient 1, was also brought with the complaint of loss of normal skin color on his body. At about the age of 5 years, small white spots were noted on his trunk, neck, and limbs; these increased in number and size with age. Patches of white hair and several areas of alopecia on the scalp were also observed. He had not had any serious illnesses, infections, operations, or complaints about his vision and hearing.

On examination, his weight was 25.5 kg (N 38.8 ± 7.3), height 128 cm (N 145.9 ± 6.1), head circumference 50.5 cm (N 53.1 ± 1.1). There were some areas of various sizes of canities and areas of alopecia 0.5 to 1 cm in diameter on the scalp. Eyebrows and eyelashes were thin with scattered whitening. He had large areas of vitiligo on his trunk, neck, and limbs (Fig. 3).

Eye examination: There were no pathological signs except increased vascular tortuosity of the fundus. His vision was normal at that time. He had chronic left otitis media. His electroencephalogram was normal. Skin test for tuberculosis and the VDRL for syphilis were negative. Cerebrospinal fluid was normal. Results of chromosome analysis were normal. Neurosensory hearing loss was found on the left by audiometry. He obtained a full scale IQ of 51 on the Wechsler Intelligence Scale for children which places his intellectual function within the moderately retarded range.

Discussion

Vogt-Koyanagi-Harada syndrome (VKH) is an uncommon disorder rarely reported in children, much less in sibs [13]. Only two instances of sibs with the VKH syndrome have been reported [1, 11]. Salus' patient was

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Fig. 1. Pedigree of patients' family

Fig. 2. Case 1. Patches of vitiligo

Fig. 3. Case 2 showing the poliosis of the scalp and the vitiligo on the trunk and extremities