Case Report

Alezzandrini syndrome: a very rare pigmentary disorder.

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Abstract

Background: Alezzandrini syndrome is a rare disorder of pigmentation. It is an acquired autoimmune disease of melanocyte characterized by segmental vitiligo, poliosis and unilateral visual and hearing impairment. An 8 years old girl presented with right sided facial leukoderma, white forelock, hypoacusis and visual disturbance for 1 and half years. Probably we were reported that type of case first time in our country.

Keywords: Alezzandrini syndrome, Vogt Koyanagi Harada syndrome, Poliosis

Introduction:

Alezzandrini syndrome is characterized by depigmented skin patches on unilateral face with white forelock and loss of acuity of vision and hearing. At first impression it mimics with a segmental vitiligo of face with poliosis.[1] The cause of this syndrome is unknown but it is thought that autoimmune processes may cause destruction of melanocyte of skin, eye and internal ear. Like Vogt-Koyanagi-Harada syndrome dimness of vision and permanent loss may occurs due to retinitis pigmentosa or retinal detachment.[2] Hypoacusis and partial or progressive ipsilateral deafness is also a feature. Medical care includes regular ophthalmological examination, hearing tests, audiometric test and management of skin lesions.

Case report:

An 8-year-old girl presented with depigmented patches on right upper two third of face with white forelock and white eyelashes on same side for one and half years. Only a few patches at outside of face e. g one is at left root of neck. She was reasonably well before that then whitish patches started to appear near her right forehead and had spread over the ophthalmic and maxillary division of trigeminal nerve. After a few months of appearance of white patches she had hearing some ringing sound off and on at right ear. For last eight months she had complaint dimness of vision and some curtain like dark objects on her right visual field.For that she consulted with an ophthalmologist and diagnosed as early retinal detachment on right eye. Audiometric test was confirmed her partial right sided sensorineuronal deafness.

Figure 1: Cutaneous features of Alezzandrini syndrome: Right sided vitiligo and poliosis

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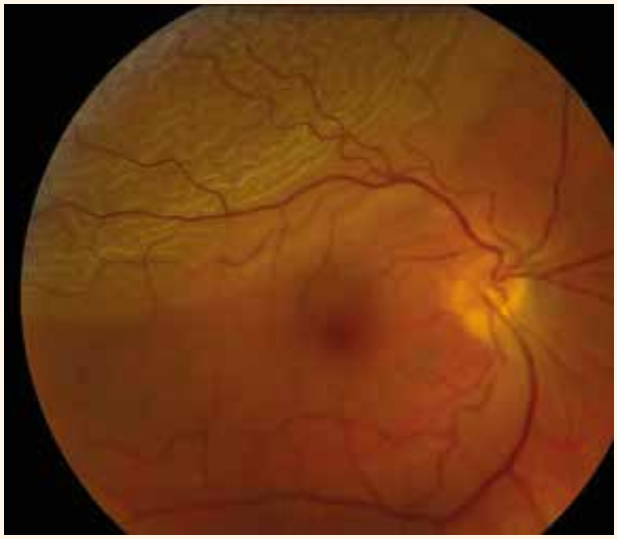

Figure 2: Retinal detachment on right eye

At initial stage of her skin lesions she was diagnosed as segmental vitiligo.Piebaldism was also a differential diagnosis. Wood's lamp study was in favor of vitiligo. Her skin lesions were not presented since birth and no other family members were affected with same kinds of lesions. For that reason piebaldism was excluded and it was assumed as an acquired disease. No history of trauma or viral infection was associated with initiation of that lesions. Vitiligo, poliosis with visual and auditory disturbance is the feature of an autoimmune disease Vogt Koyanagi Harada syndrome. The features of VKH syndrome is usually bilateral and generalized but it was localized and unilateral in that girl. For above reasons that patient was diagnosed as a case of Alezzandrini syndrome.

There was no specific treatment option in Alezzandrini syndrome. The features of that girl was progressive. The patient had suggested for regular ophthalmological and hearing monitoring and support.

Discussion:

Alezzandrini syndrome is an acquired autoimmune disorder of neural crest derivative specially the melanocyte. The first case was described by Casala and Alezzandrini in 1959. They found a case with vitiligo, poliosis hyperacusis and unilateral retinitis pigmentosa.[3]The similar disorder of melanocytes in skin, internal ear and eyes was well known as Vogt Koyanagi and Harada syndrome where the features was bilateral.[4] Alezzandrini syndrome is a very rare disorder that only the sixth case was reported by Andrade & Pithon in 2011.[5]Around 10 cases were reported worldwide up to the date.

The exact etiology of this syndrome is unknown, but one hypothesis is an autoimmune disease targeting melanocytes and other neural crest derivatives. Patients present with unilateral depigmentation of the facial hair and skin and ipsilateral loss of visual and auditory acuity.[6] Ophthalmological examination and audiometry is needed for confirmation and monitoring of cases. There is no specific treatment option for Alezzandrini syndrome. A combined effort of dermatologists, ophthalmologist and otolaryngologist will be supportive for patient for better life style. The syndrome progresses over years.

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