Scleritis, poliosis and dermatitis; A new ophthalmologic syndrome?

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Abstract
Aggregation of some sign and symptoms from different organ system are important issues in medicine. Scleritis, vitiligo, poliosis, dermatitis and multiple sclerosis-like neurologic feature are features from a single pathophysiology. We describe for the first time a 23-years-old male with history of whitish hair discoloration and hypopigmented skin lesions, severe burning sensation all over the body and painful red eye. We proposed a new syndrome which has not been reported yet that merits further attention in clinical practice.

Keywords: poliosis, Scleritis, hypopigmentation, dermatitis, multiple sclerosis-like, VKH-like, vitiligo.

INTRODUCTION
Scleritis is a disease that affects the white outer coating of the eye, known as the sclera. The disease is often associated with other diseases of the body, such as Wegener's granulomatosis or rheumatoid arthritis. Co-incidence of eye, skin and neurologic findings are reported in several syndromes. Behcet's syndrome, for example is manifested as constellation of uveitis, recurrent oral (and/ or genital) aphthi, musculoskeletal and neurologic symptoms. Behcet disease is primarily considered as an autoinflammation of the blood vessels (1). Vogt-Koyanagi-Harada also manifests as hypopigmentation, uveitis and neurologic symptoms with separate clinical outcomes (2). Most of these diseases are immune-mediated. The detection of systemic Vasculitis in cases with scleritis is associated with poor prognosis (3). The most frequent infectious cause of scleritis is herpes zoster (4). Classification of these syndromes could specify outcome and better management. Herein we describe a 23 years-old man with new association of scleritis, hypopigmentation, dermatitis and multiple sclerosis-like neurologic manifestations. We search Medline and Google Scholar for similar entity but no matched aggregation was found.

CASE REPORT
A 23-year-old male with history of whitish hair discoloration and hypopigmented skin lesions on left side of his face since 3 years ago, severe burning sensation all over the body since 3 months ago which was aggravated with high temperature conditions and painful red eye 6 months ago which has been managed initially as conjunctivitis. The patient underwent a systematic evaluation, including clinical history; general, neurological and ophthalmologic examinations. Skin, teeth, nails and hair were examined for defects. Routine laboratory tests for blood, urine, were performed. Special examinations pertaining to mental performance, biochemistry, imaging and electrodagnostic studies, and skin biopsy were also performed.

Patient's description and medical records of dermatology visit were consistent with vitiligo on left lateral side of his face and neck which was managed with topical corticosteroid remedies. In eye exam, he showed 10/10 vision, with normal eye movements. In slit lamp examination, conjunctival swelling and redness and marked congestion of scleral vessels without any discharge of his left eye consistent with scleritis was evident. No inflammation was found in anterior chamber and pupillary reflex was normal. Intra-ocular pressures were 14 and 15 mm of Hg for right and left eye respectively. Both lenses were clear and no abnormality was found on posterior segment after full pupillary dilatation. Examination of his hair showed hypopigmented whitish hair (Figure 1) and eczematoid skin change on his right arm. (Figure 2) His neurologic examination revealed exaggerated
deep tendon reflexes in patellar and Achilles tendons, unilateral mild clonus and positive Babinski reflex. Routine lab investigations and visual evoked potential study and brain MRI (without gadolinium) showed normal results. Patient was treated with oral prednisolone 40 mg/day, topical steroid and systemic non-steroidal anti-inflammatory drugs. Rapid clinical improvement ensued.

DISCUSSION
Scleritis (inflammatory condition of sclera) is classified to four types: diffuse, nodular, necrotizing and posterior scleritis. Clinical manifestations include congestion of sclera and conjunctiva, boring globe pain, photophobia alone or with lacrimation and in severe cases vision impairment. The diagnosis is based on slit lamp examination. In many cases, scleritis is in the context of overt or occult systemic conditions. In one study on 29 patients with scleritis, 55.7% had underlying systemic condition, of them 34% with rheumatologic and 20% with infectious (brucellosis, tuberculosis, chicken pox and mumps) diseases (5). Among rheumatologic conditions, rheumatoid arthritis, systemic lupus erythematosus, polychondritis, polyarteritis nodusa, granulomatosis with polyangitis (WG) and ankylosing spondylitis are the leading causes of scleritis. Scleritis also has been reported as a postoperative complication (6). Inflammation of sclera (Scleritis) is usually as a result of immunologic response and type III and IV immune responses are involved in pathophysiology of scleritis. Addressing to the base of immune aberrancy, other features of this derangement is frequently seen in association with scleritis. Vitiligo is one the immune mediated conditions with lack of pigmentation of the skin. Poliosis or loss of normal pigmentation of hair is also frequently associated with known immunologic syndromes such as Vogt-Koyanagi-Harada (VKH). In nervous system the prototype of immune mediated demyelination is multiple sclerosis (MS). In the present case albeit no demonstrable central nervous system finding were observed in brain MRI, but subtle and mild upper motor neuron lesions is highly the case with mentioned clinical findings.

Getting together, this entity implies to an abnormal immune response targeting different organ systems. Yeh et al reported association of scleritis with systemic manifestations of vitiligo, psoriatic arthritis, thyroiditis, erythema nodusum, and Sjogren's syndrome. In this study abnormal increase of natural killer T cells was observed (7). In another study, Koudu et al has reported association of posterior scleritis with VKH (8). Lin et al showed that in more than half of patients suffering from scleritis associated with rheumatoid factor positivity, frank rheumatoid arthritis developed after short period of time (9). Considering high association of scleritis and systemic diseases, a thorough medical history, physical examination and aggressive anti-inflammatory treatment is advised (10, 11). This could be the first report of constellation of scleritis, vitiligo, poliosis and MS-like presentation in a patient. So, if other observations proved similar presentation, it definitely could be considered as a new and independent entity of "Manaviat-Owlia syndrome".

REFERENCES
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