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Case Report

Congenital Atrichia with Recurrent Episcleritis-A Rare Case Report

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Abstract

Congenital atrichia is a rare disorder and is often associated with various systemic diseases. Ocular involvement has also been reported. We report a rare case of congenital atrichia with recurrent episcleritis. To the best of our knowledge and after an extensive internet search, this is probably the first reported case of congenital atrichia with episcleritis.The association may be a co-incidental finding or may have a broader perspective. Hence, further studies are warranted.

Keywords: Renal; Nephrotic syndrome; Keratitis; Congenital nystagmus; Cone-rod dysfunction

1. Case Study

A 35-year-old female patient presented to the department of ophthalmology with a history of redness, lacrimation and photophobia in the right eye off and on for the past one year (Figure 1). There was no other ocular complaint. There was no other significant medical, surgical, family, traumatic or drug abuse history. Ocular examination was carried out and her visual acuity was 6/6 in both the eyes; pupillary reactions, ocular movements, colour vision, fundus and intraocular pressure were normal bilaterally. Slit lamp and torch light examination revealed simple episcleritis in the right eye.

There were absence of eyebrows and eyelashes on examination. On further questioning the patient, she removed her cap to reveal the total absence of hairs on the scalp. She further stated that there was a complete absence of hairs since birth and dermatological consultation had revealed congenital atrichia. Rest of her general physical and

systemic examination was normal. For episcleritis, the patient was started on tablet naproxen and fluorometholone eye drops. Blood investigations to rule out connective tissue disorders were within normal levels. To the best of our knowledge and after an extensive internet search, this is probably the first reported case of association between episcleritis and congenital atrichia.

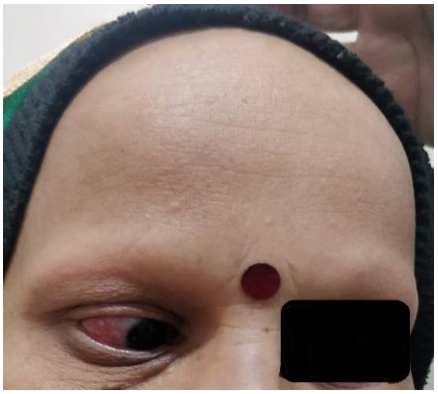

Figure 1: Patient with a history of redness, lacrimation and photophobia in the right eye.

2. Discussion

Congenital atrichia is a rare disease causing irreversible alopecia which is inherited autosomal recessively. There is complete irreversible hair loss seen soon after birth. Situs inversus and mesocardia has also been reported to be associated with this disease. Other associations include Moynahan's syndrome (characterized by seizures with mental retardation), hidrotic ectodermal dysplasia (palmoplantar keratoderma, thickened nails) and aging syndromes [1]. Loss of scalp hair is seen between one to six months of age, after which no growth occurs. There may also be sparse or absent hairs on the eyebrow, eyelash, and body. Differential diagnosis include vitamin D dependant rickets, congenital alopecia universalis, and ectodermal dysplasia [2]. IFAP (Ichthyosis follicularis with alopecia and photophobia) syndrome has also been reported in literature where patient presents with a triad of striking alopecia, photophobia and generalized cutaneous 'thorn-like' projections [3].

Atrichia has also been reported to be associated with anomalies of the face, nails, cartilage, speech, respiratory tract, digits, and oral cavity. Renal involvement in the form of nephrotic syndrome and glucosuria has also been reported. Ocular involvement in the form of congenital cataracts, microphtalmia, keratitis, congenital nystagmus, cone-rod dysfunction, and high myopia has been reported [4]. Episcleritis is the inflammation of the episclera (a thin, loose, highly vascular connective tissue layer between the conjunctive and sclera). It can be idiopathic, or it can be associated with connective tissue disorders like rheumatoid arthritis, scleroderma and systemic lupus erythematosus. Watson and Hayreh classified episcleritis into simple and nodular. Most patients have intermittent bouts of moderate or severe inflammation at intervals of 1–3 months. Symptoms include acute onset of redness, lacrimation, and photophobia. It commonly affects a single quadrant in one eye. Bilateral involvement suggests underlying systemic disease. Systemic non steroidal anti-inflammatory drugs (NSAIDs) and topical steroids are used in its treatment [5].

Conflicts of Interest

The authors declare that they have no competing interests.

Financial Disclosure

The authors have no proprietary or commercial interest in any material discussed in this article.

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