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Ross syndrome

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ABSTRACT

Ross syndrome is a rare disease consisting of a triad of Holmes adie pupil, hyporeflexia of deep tendon reflexes and segmental anhydrosis. There are about 40 case reports of the same worldwide. In this case report we describe on such case.

KEYWORDS Ross syndrome, Holmes adie pupil, anhidrosis, hyperhidrosis

Introduction

Ross syndrome is a rare disorder with only about 40 case reports of the same worldwide. It consists of a triad of findings- holmes adie pupil, segmental anhidrosis and hyporeflexia or areflexia. In this case report we report one such patient.

Case description

A 30 year old gentleman presented to the medical outpatient facility with difficulty in seeing in the bright light since a year with associated sweating in only one half of the body since 2 years. He had no other systemic symptoms.

On examination, he had a holmes adie pupillary response with dilated pupil in response to light on the left side (figure 2). Accommodation reflex was well preserved. His deep tendon reflexes were hyporeflexic. Segmental compensatory hyperhidrosis was seen (Figure 2) in the region of the T3-T12 dermatomes. Rest of the systemic examination was normal.

Extensive laboratory evaluation was carried out to rule out other features of vasculitis, sarcoidosis, CMV infection, autoimmune features and syphilis. However there was no evidence of any of these on serology, PCR testing, MRI brain with spinal cord and cervical spine imaging or CT imaging of the thorax and abdomen.

Discussion

Ross syndrome is a rare entity with holmes adie pupil and segmental anhidrosis usually accompanied by compensatory hyperhidrosis on the other side. It forms a spectrum of disorders affecting the cholinergic neurons along with the harlequin syndrome and holmes adie syndrome.

The Harlequin syndrome is characterised by segmental anhidrosis without the pupillary anomalies while the holmes adie syndrome is characterised by the pupillary abnormalities without the segmental anhidrosis. The association between the two of them was first made by Ross in the year 1958. They are believed to be the same spectrum of disorder.[ii]

It is thought to be a degenerative disorder of the postganglionic cholinergic neurons. The pupillary findings have been thought to be due to denervation of neurons between the ciliary ganglion and sphincter pupillae.[iii] In the anhidrotic areas there is usually are marked loss of nerve fibres. Aetiology is unknown. It may be associated with evidence of other autonomic dysfunction including alterations in heart rate. Course is usually slowly progressive with gradual expansion of the dermatome involved even upto 50 years of age.[iv]

Treatment is with videoassisted thoracoscopic sympathetectomy and injection of botulism toxin to address the issues of hyperhidrosis in that dermatome.[v]

Figure 1: Right sided pupil constricted with left sided dilated pupil in response to light

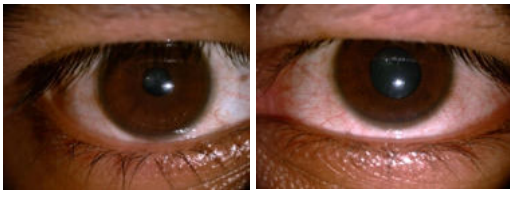
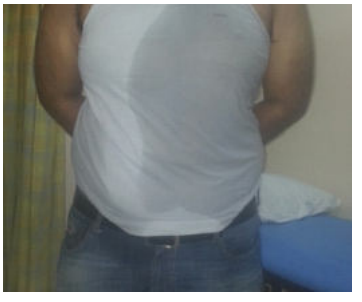


Figure 2: Segmental anhydrosis on one half of the body with compensatory lhyperhydrosis on the other half of the body



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