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Case Report

Ross syndrome: a case report

Manoj Kr. Mahata[1]*, Saikat Ghosh[2], K. C. Ghosh[3], R. Bhattacharyya[3], G. P. Mondal[3]

[1]Department of Neurosurgery, Hyogo College of Medicine, Hyogo, Japan
[2]Department of Neurology, Manorama Hospitex, Nadia, West Bengal, India
[3]Department of Neurology, CNMC, Kolkata, West Bengal, India

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***Correspondence:**
Dr. Manoj Kr. Mahata,
E-mail: manojmahata85@gmail.com

ABSTRACT

Ross syndrome is a rare partial dysautonomic syndrome of unknown aetiology, characterized by segmental hypo/anhidrosis associated with Holmes-Adie syndrome (tonic pupil and hypo/areflexia). The hypohydrosis or anhydrosis is patchy initially, later it becomes segmental or diffuse. This is due to affection of postganglionic cholinergic parasympathetic and sympathetic fibers involvement. There are a very few cases (approximately 50) have been reported in the literature since its original description. Author report a 22 years old male with classical features of Ross syndrome.

Keywords: Anhidrosis, Areflexia, Ross syndrome

INTRODUCTION

Ross syndrome is a rare partial dysautonomic syndrome of unknown aetiology, characterized by a one-sided or bilateral anhidrosis associated with Holmes-Adie syndrome (tonic pupil and hypo/areflexia).[1] It is a spectrum disorder with Harlequin and Holmes Adie's syndrome in two ends, and Ross syndrome is the combination of two.[1] Ross syndrome may have an unpredictable course and its causation may be due to various factors like autoimmunity, developmental origin, viral infections etc.[2-6] Since its first description in 1958, approximately 50 cases have been described in literature and only 3 have been reported from India so far.

CASE REPORT

A 22-year-old male, with no known comorbidities, presented with heat intolerance and absent sweating on the right half of the body since childhood. As the patient worked in a leather industry where the ambient temperature is high, he felt episodes of intense heat and intolerance especially on the right half of body although there was no history of previous hospitalization or any application of topical medicines. There was no history of trauma to the spine, syncopal attacks, weight loss, pain abdomen, speech or memory deficits or any evidence of sphincter dysfunction. General clinical examination was normal. Blood pressure readings were normal and not suggestive of postural hypotension. Cutaneous examination of the right side of the face and upper and lower limbs revealed absent sweating (Figure 1). Higher mental functions were normal. Pupils of both the eyes were sluggish in their reaction to light and on adding 0.125% pilocarpine drops, there was constriction of both the pupils (Holmes Adie pupil). Stretch reflexes in all 4 limbs were absent. Other systemic examinations including the central nervous system and spine were normal. Routine investigations like hemogram, urine examination and thyroid profile were within normal

limits. Veneral disease and research laboratory (VDRL) test was nonreactive. Chest X-ray and magnetic resonance imaging, (MRI) of brain, cervical, thoracic and lumbosacral spine detected no abnormality. Nerve conduction study (NCS) of all 4 limbs was normal. Histopathological examination from the anhidrotic area showed sparse to absent eccrine sweat glands.

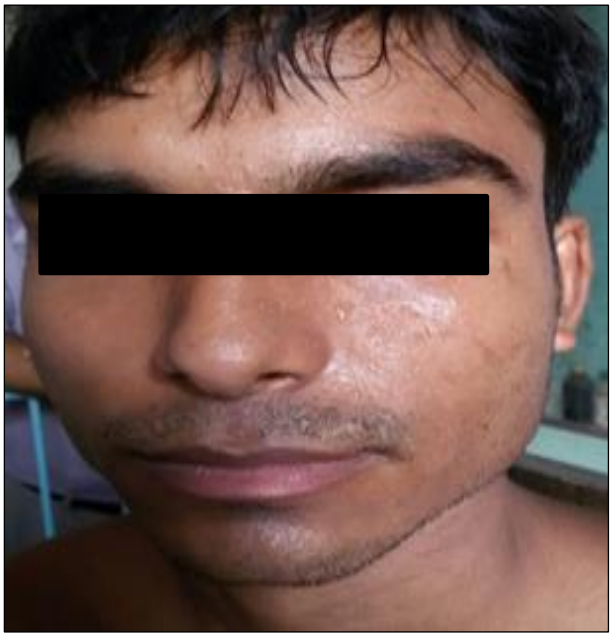

Figure 1: Anhidrosis of right face.

DISCUSSION

Ross syndrome is a rare disorder of sweating comprising of widespread hypo/anhidrosis combined with patchy compensatory hyperhidrosis associated with areflexia and tonic pupil (Holmes Adie syndrome). It affects both males and females with age of onset ranging from 3 to 50 years. It is considered as the expression of an unknown injury to the peripheral autonomic nervous system. Its exact pathogenesis is unknown. A wide overlap has been suggested between Ross syndrome, Holmes-Adie syndrome and more widespread autonomic disease.[7] When anhidrosis is extensive, the remaining areas of the functioning eccrine glands may show compensatory hyperhidrosis. The compensatory hyperhidrosis may be striking and severe enough to require therapy, although eventually it may be lost as complete anhidrosis develops. The hyperhidrosis could be compensatory or due to early loss of cholinergic M2 inhibitor presynaptic autoreceptors. The sweating disturbance could result from a lesion of the sympathetic ganglion cells or their post-ganglionic projections.[8] The hypothesis of a degenerative mechanism has recently been reported involving sudomotor fibres and other skin autonomic nerve fibres (innervating notably blood vessels and arrector pilorum muscles).[1] More recently, a direct infectious damage to autonomic nerves by an acute cytomegalo virus (CMV) infection has been hypothesized in an isolated case report.[5]

CONCLUSION

Although most cases of Ross syndrome are detected by dermatologists, clinical suspicion of the same must be made from a neurologist's point of view when a patient of anhidrosis is encountered.

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