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Case Report

An interesting case of anhidrosis

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ABSTRACT

Ross syndrome is a rare syndrome of autonomic dysfunction which is characterized by segmental hypohidrosis/ anhidrosis, Adie tonic pupil and hyporeflexia/areflexia. Exact etiology is not known. It was first described in 1958 and approximately 50 cases have been described so far globally. We hereby report a case of segmental anhidrosis of left face, left upper limb and left side of chest with 3-year duration in a 26-year-old man who also had Adie's tonic pupil in right eye and global areflexia on examination.

Keywords: Autonomic dysfunction, Adie's tonic pupil, Anhidrosis, Ross syndrome

INTRODUCTION

Ross syndrome is a rare syndrome of unknown etiology. This syndrome was first described in 1958 by Ross and so far, approximately 50 cases have been reported.[1] This syndrome is characterized by autonomic dysfunction with a clinical trial of segmental hypohidrosis/anhidrosis, Adie tonic pupil and hyporeflexia/areflexia. Dysfunction of postganglionic cholinergic fibres causes pupil and sweating abnormality. Dorsal root ganglion degeneration and loss of spinal interneuron cause depressed deep tendon reflexes.[2]

CASE REPORT

A 26-year-old man presented to our outpatient department, with history of absent sweating in left face, left upper limb and left side of chest for past 3 years which was associated with excessive sweating in right chest and right upper limb. He also complained of severe burning sensation in left upper limb till elbow which used to get aggravated in hot climate. There was no prior history suggestive of rheumatological disease, trauma or stroke. On examination of pupils, he had right tonic pupil with light near dissociation. He had diffuse areflexia. Sweating was absent in left face, left upper limb and left side of chest. Ptosis was absent and rest of neurological examination was normal. His magnetic resonance imaging of brain and magnetic resonance imaging of cervical spine with whole spine screening, chest X-ray, ultrasonography of abdomen and pelvis were normal. His blood investigations like fasting and postprandial blood sugar, fasting lipid profile, complete blood count, renal and hepatic parameters, thyroid function test, erythrocyte sedimentation rate were normal. Antinuclear antibody testing by enzyme linked immunosorbent assay, split skin smear for acid fast bacilli and veneral disease research laboratory test for syphilis (VDRL) were negative.

Nerve conduction study of all 4 limbs were normal. Sympathetic skin response was negative in left upper limb and lower limb, and normally obtained in right upper limb and lower limbs. Bedside autonomic function tests were normal. Clinical diagnosis of Ross syndrome was made on the basis of clinical trial of segmental anhidrosis, Adie tonic pupil and areflexia.

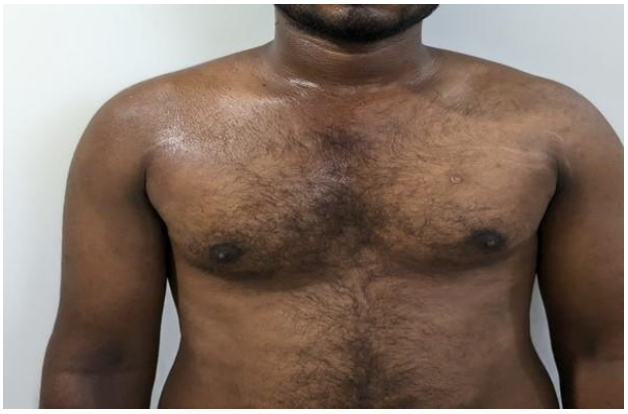

Figure 1: Anhidrosis in left chest and left upper limb seen. Hyperhidrosis seen in right side of chest and right upper limb.

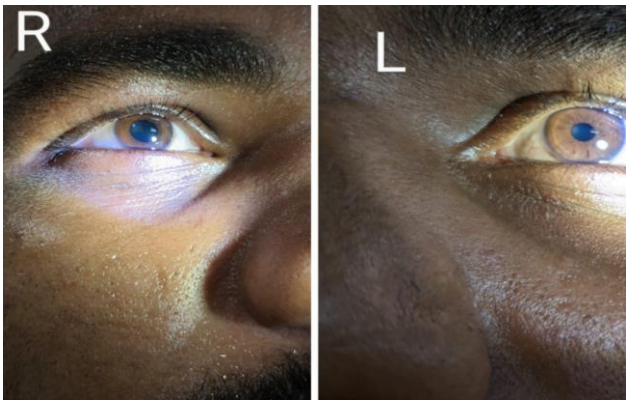

Figure 2: Right tonic pupil present with no reaction to light; left side pupil normally reacting to light present.

DISCUSSION

Ross syndrome is a rare clinical entity caused due to autonomic dysfunction. Mean age at time of diagnosis was 36 years with slight female predominance. Dysfunction of sympathetic ganglion cells/post ganglionic projections could be the reason for loss of sweating. Abnormality in dorsal root ganglion/spinal interneuron loss may cause depressed deep tendon reflex.[3] Adie's pupil was said to be due to denervation of postganglionic cholinergic fibers between ciliary ganglia and iris sphincter muscle.[4] Holmes-Adie syndrome mimics this condition, but sweating abnormality is not seen in Holmes-Adie syndrome. Possibility of autoimmunity, genetic factors, and developmental abnormality, has been suggested in the etiopathogenesis of this syndrome, but the exact etiology remains unknown.[5] Anhidrosis develops slowly over a time and can be accompanied with area of compensatory hyperhidrosis. Sometimes other autonomic disturbance like Horner's syndrome, cardiac dysautonomia may be seen.[4] There is no definitive treatment. Iontophoresis, botulinum injection and topic glycopyrrolate were found to be useful in hyperhidrosis management. For heat intolerance, wet clothing during summer and during physical exertion can be tried.[6]

CONCLUSION

Ross syndrome is a rare disorder with the clinical trial of segmental hypohidrosis/anhidrosis, Adie's tonic pupil and hyporeflexia/areflexia, the exact etiology of which is not known. About 50 cases of Ross syndrome were reported globally and very few cases have been reported from India.

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