*IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*
*e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 23, Issue 1 Ser.4 (January. 2024), PP 40-41*
*www.iosrjournals.org*

# Ross Syndrome : A Case Report

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***Abstract***
*Ross syndrome is a rare disease entity caused due to partial autonomic dysfunction. It is represented by a triad of areflexia or hyporeflexia, Adie's tonic pupil and hypohydrosis or anhydrosis. We present case of a 46 year old male with complaint of excessive sweating over left side of trunk since 2 years. Examination revealed hyporeflexia and adie's tonic pupil. Very few cases (~50) have been reported about this disorder till date.*
***Key words:*** *Ross syndrome, areflexia or hyporeflexia, anhydrosis or hypohydrosis, adie's tonic pupil*

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Date of Submission: 05-01-2024 Date of Acceptance: 15-01-2024

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## I. Introduction

Ross syndrome is a rare partial dysautonomic syndrome of unknown aetiology, characterized by a one-sided or bilateral anhydrosis associated with Holmes-Adie syndrome (tonic pupil and hypo/areflexia). 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 has been attributed to a large number of factors such as autonomic denervation, autoimmunity, developmental origin, viral infections, and genetic factors.[2-6] There has been paucity of reports of this syndrome till date, compelling us to report this case of middle aged man from Andhra Pradesh.

## II. Case report

This 46 year old male, known case of hypertension since 1.5 years presented with complaints of excessive sweating on left side of trunk. It occurred daily, even at rest. As patient was driver by occupation, he had no complaints as such except for increased sweating. There was no history of any trauma to spine, low backache, bowel bladder dysfunction, syncopal attacks, headache, palpitations, chest pain or dyspnea. There were no similar complaints in family. On examination vitals were stable with no evidence of orthostatic hypotension. There was increased sweating in T4-T9 dermatomes of trunk- both front and back on left side only. On ophthamological examination right eye pupil was 4mm (Figure 1) and was sluggishly reactive to light while left eye pupil was 2mm. Both pupils constricted on adding 0.125% pilocarpine drops. Tone and power were bilaterally normal. Deep tendon reflexes in all four limbs were 1+ (diminished). Lab investigations such as hemogram, thyroid profile, renal function test, liver function test and random blood sugar were normal. ANA, VDRL, HIV, HbsAg and HCV were negative. Chest X-ray and USG abdomen showed no abnormality. NCS all 4 limbs and MRI brain with whole spine screen were also normal. Since patient did not complaint of any discomfort except excessive sweating, he was started on Tab Glycopyrrolate 2 mg BD and was asked to follow up to assess the response and it was followed by decrease in his symptoms.

## III. Discussion

The findings of hyperhidrosis, hyporeflexia and Adie's tonic pupil helped to substantiate the diagnosis of Ross syndrome in this patient. 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.[7] Defects in thermoregulation along with anhidrosis can lead to periodic hyperthermia in such patients that could be life threatening. Damage to postganglionic sympathetic fibers of the sweat glands is responsible for segmental and progressive hypohidrosis.[8] 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.[9]

The loss of DTR is due to dorsal root ganglionic degeneration and spinal inter neuronal loss.6 The pathophysiology of the tonic pupil is due to damage to parasympathetic cholinergic fibres between iris and

ciliary ganglion6and resultant cholinergic supersentivity.[10] Management depends on symptomatology. If hyperhidrosis is the chief complaint botulinum toxin,[11] iontophoresis,[12]aluminum chloride, 0.5% glycopyrrolate,[13] sympathetic thoracotomy[14] and systemic anticholinergics have been advied with varying results. Management of hypohidrosis depends on nonpharmacological measures like avoidance of hot climate and wearing wet clothes during exertional activities. One case report of ANA positivity[15] and subsequent improvement with IVIg and one case report of cytomegalovirus positivity[16] and partial improvement with resolution of infection has been documented.

## IV. Conclusion

Being a rare disease entity Ross syndrome is often left undiagnosed by the practicing clinicians. Based on symptomatology patients could consult a physician, dermatologist, ophthalmologist or neurologist. Hence, awareness about this entity is needed. Although treatment options are limited, but correct diagnosis may help to alleviate anxiety amongst the patients and may help them to discover their own coping strategies timely.

**Declaration of patient consent:** The patient has given consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity but anonymity cannot be guaranteed.

**Financial support and sponsorship:** Nil

**Conflicts of Interest:** There are no conflicts of interest

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**Figure 1: shows Adie's tonic pupil**