

# Unique Ailment, Unusual Affliction: Ross Syndrome

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## Abstract

Ross syndrome is a rare disorder characterized by Adie's syndrome and segmental anhidrosis. It was first described in 1958 by Ross. There is involvement of the autonomic innervation which causes loss of sweat thereby impairing heat dissipation. This can be attributed to the affection of postganglionic cholinergic parasympathetic and sympathetic fibers to the sweat glands. We present a variant case of Ross syndrome in which the patient presented with anhidrosis of left half of the body. Thermal imaging revealed hemi anhidrosis and ophthalmic examination showed Adie's pupil. Neurological examination elicited hyporeflexia of deep tendon reflexes of left side. 3T magnetic resonance imaging of sympathetic plexus showed no abnormality. This case is presented as a rare variant of Ross syndrome.

**Key words:** Adie's pupil, Anhidrosis, Ross, Thermal imaging

## INTRODUCTION

Ross syndrome is a rare disorder characterized by Adie's syndrome and segmental anhidrosis. It was first described in 1958 by Alexander Ross. Less than 50 cases have been reported till date.<sup>1</sup> In Ross syndrome, there is involvement of the autonomic innervation which causes loss of sweat thereby impairing heat dissipation. This can be attributed to the defect of postganglionic cholinergic parasympathetic and sympathetic fibers to the sweat glands. Adie's syndrome is characterized by Adie's pupil (tonic pupil with slow reaction to light) and absent or diminished deep tendon reflexes.<sup>2</sup> We present a rare variant of Ross syndrome in a young male.

## CASE REPORT

A 25 years male presented in our tertiary care center with complaints of loss of sweating in the left half of the body

noticed 3 months back. He noticed left half of the body was dry while the other half was sweating. There was no history of invasive procedures or trauma. There was no other significant positive history. He was an occasional alcoholic for 1.5 years.

On examination, vitals were stable. There was no evidence of postural blood pressure changes. The individual was subjected to mild exercise (brisk walk) in which anhidrosis of left half of the body was appreciated as shown in Figure 1. Ocular examination revealed visual acuity of 6/6 in the right eye and 6/9 in the left eye. Anterior segment examination showed no abnormalities in the right eye. Left eye showed dilated pupils of 5 mm with no reaction to both direct and indirect light reflex (Figures 2 and 3). Pilocarpine drops were instilled which confirmed Adie's tonic pupil of left eye. Posterior segment evaluation was normal.

Routine blood investigations were within normal limits. 3T magnetic resonance imaging of sympathetic plexus were done and showed no abnormalities. Thermal infrared imaging showed a significant rise in temperature of left half of the body due to loss of heat dissipation by sweating (Figures 4-6). Hence, we confirmed the diagnosis of Ross syndrome.

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Figure 1: Anhidrosis of left half of body after exercise

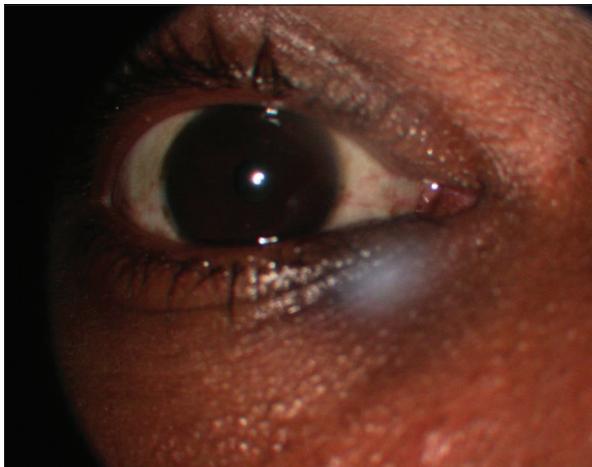


Figure 2: Normal pupil of right eye

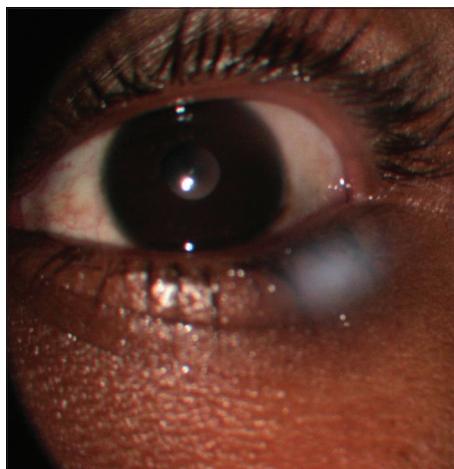


Figure 3: Dilated and nonreacting pupil of left eye

## DISCUSSION

Ross syndrome is a rare neurological disorder affecting both sexes with slight female predominance.<sup>3</sup> Previous literature described Ross syndrome as segmental anhidrosis with Adie's syndrome. In our case, there is anhidrosis of left side of face, neck, trunk extending till the medial aspect of the thigh along with tonic pupil and decreased deep tendon reflexes of same side. Decrease in deep

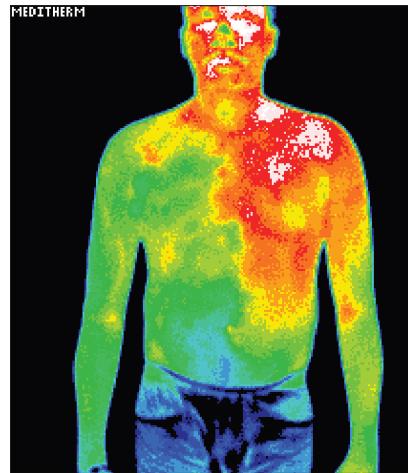


Figure 4: Thermal infrared imaging showing significant rise in temperature

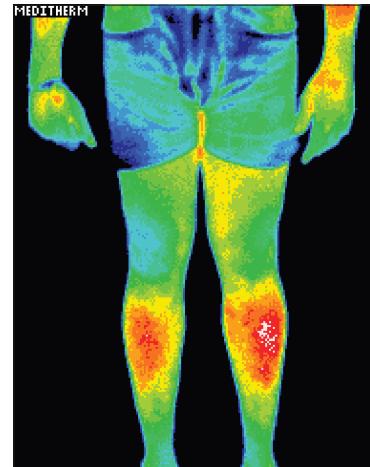


Figure 5: Thermal infrared imaging showing significant rise in temperature

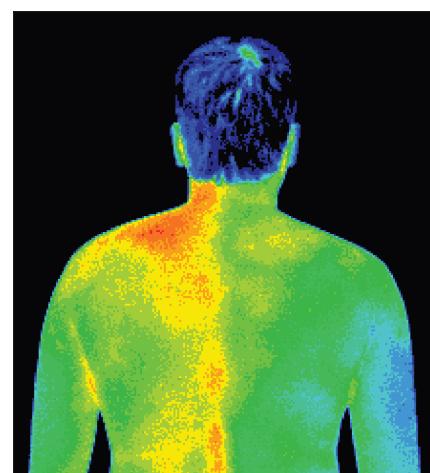


Figure 6: Thermal infrared imaging showing significant rise in temperature

tendon reflexes is due to damage of the dorsal root ganglia.<sup>4</sup> Anhidrosis is due to defective postganglionic cholinergic parasympathetic and sympathetic fibers to the

sweat glands whereas Adie's pupil results due to damage of the parasympathetic cholinergic fibers between the iris and ciliary ganglion.<sup>5</sup> Harlequin syndrome is a condition where there is asymmetric sweating and flushing of the upper thoracic region of the chest, neck, and face.<sup>6</sup> We excluded the diagnosis of Harlequin as there was pupillary involvement in our patient and decreased deep tendon reflexes. Defects in thermoregulation with anhidrosis can lead to hyperthermia of the affected side which can be life threatening.<sup>7</sup> Hyperthermia can be managed to some extent by wearing wet clothing during physical activity.<sup>8</sup> This case is presented for its rare variant of Ross syndrome.

## CONCLUSION

Ross syndrome is a rare neurological disorder affecting both sexes with slight female predominance. Previous literature described Ross syndrome as segmental anhidrosis with Adie's syndrome

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