

A RARE CASE REPORT OF ROSS SYNDROME

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ABSTRACT

Ross syndrome is a degenerative peripheral nervous system disorder defined by the following triad: unilateral or bilateral segmental anhidrosis, hyporeflexia of deep tendon reflexes and Adie's tonic pupils. The most disturbing symptom is segmental compensatory hyperhidrosis. It has only occasionally been reported in the dermatological literature. We present a 27-year-old man who developed the characteristic triad of Ross syndrome since childhood with a positive family history of Adie's tonic pupil in his brother.

Keywords: Ross Syndrome, Adie's Tonic Pupil, Hypohidrosis

INTRODUCTION

Ross syndrome is a rare disorder of sweating comprising of widespread hypohidrosis combined with patchy compensatory hyperhidrosis associated with is flexia and tonic pupil (Coulson, 2004). It affects both males and females with age of onset ranging from 3 to 50 years. Patients with Holmes-Adie syndrome often show asymptomatic changes in sweating. This anhidrosis may be localized or widespread. When anhidrosis is extensive, the remaining areas of the functioning eccrine glands may show compensatory hyperhidrosis (Ross, 1958). Cardiac sympathetic denervation that may be asymptomatic has also been shown to develop. The compensatory hyperhidrosis may be striking and severe enough to require therapy, although eventually it may be lost as complete anhidrosis develops (Lucy *et al.*, 1967). It is thought that in anhidrotic areas, there is severe loss of sudomotor fibers. There is only a slender network of protein gene product immunoreactive fibers. These fibers do not have receptors for vasoactive intestinal peptide or dopamine- β -hydroxylase axons (Nolano *et al.*, 2006). The hyperhidrosis could be compensatory or due to early loss of cholinergic inhibitor presynaptic autoreceptors (Ballester-Diez *et al.*, 2005). Ross syndrome is a rare disorder of sweating associated with depressed or absent deep tendon reflexes and tonic pupil due to selective degeneration of sympathetic pathways. About 40 cases of Ross syndrome have been reported so far. There is no effective therapeutic management for this condition. Heat intolerance may be managed by wearing wet clothing during physical activity in order to prevent hyperthermia and hyperhidrosis to some extent by iontophoresis (Reinauer *et al.*, 1993) and botulinum toxin injection (Bergmann *et al.*, 1998). Recently, topical glycopyrrolate was found to be safe and effective in controlling compensatory hyperhidrosis (Bajaj *et al.*, 2006).

CASES

A 27 year old male, presented to the Dermatology OPD with the complaints of loss of sweating over left half of face and right side of the body since the age of sixteen. There was also complaint of increased sweating on the other side. Patient noticed of generalized burning sensation on sun exposure and generalized dry skin on the arms and legs since childhood. Interestingly, he also noted a difference in size of the pupil at that time. This was associated with photophobia. There was history of increased thirst and frequency of micturition. There was no history suggestive of decreased salivary secretion, decreased tear secretion, decreased growth of hair or nails, except on the face. Patient underwent appendectomy 3 months back. He was not a smoker but consumed alcohol once a week. Patient had not taken any treatment for his symptoms. There was no similar complaint in the family except an unequal pupillary size in his brother.

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A general clinical examination was within normal limits. There was no significant difference in the blood pressure in upper and lower limbs in supine and prone positions. Cutaneous examination showed hyperhidrosis on the right side of face [Figure 1]. There was absence of sweating on the left side with sparse hair on the cheek [Figures 2 & 3]. On the trunk, anhidrosis was observed on the right side and hyperhidrosis on the left side [Figure 4]. There was absence of sweating on both palms and soles with fissuring on both soles. There was localized ichthyosis over both lower limbs.

Ophthalmic Examination: Pupillary examination showed anisocoria, with the left pupil larger than the right [Figures 5 & 6]. Bilateral - visual acuity, colour vision and external ocular movements were normal. Ptosis was not observed. The left pupil was mydriatic. Consensual reaction of the right pupil was normal and absent in left pupil. Both pupils, however, reacted well to accommodation, which meant that near reaction was much better than light reaction of the left pupil. These features were in accordance with a tonic pupil in the left side. Perimetry, fundus examination, visual fields of both eyes were within normal limits. Visual acuity was 6/6 in both eyes. Examination of eyes suggested the presence of Adie's pupil in the left eye [Figure 7]. Examination of spine was normal. There was absence of knee and ankle joint reflexes in both sides. Routine investigations like hemogram, urine examination and thyroid profile were within normal limits. VDRL test was nonreactive. Chest X-ray and radiographs of the cervical, thoracic and lumbosacral spine detected no abnormality. Nerve conduction studies were within normal limits. Histopathological examination from hyperhidrotic and anhidrotic patches were taken which revealed absence of sweat glands on the right side of trunk and normal sweat glands on the left side [Figures 8 & 9].

DISCUSSION

The diagnosis of Ross syndrome was based on the clinical symptoms, neurologic signs like:

- a. Segmental anhidrosis with compensatory hyperhidrosis,
- b. Absence of deep tendon reflexes especially knee and ankle joints
- c. Pupillary abnormality suggestive of Adie's pupil.

We considered the differential diagnosis like Holmes-Adie syndrome, Horner's syndrome and hereditary sensory autonomic neuropathy.

1. Adie's syndrome or Holmes-Adie's syndrome or Adie's tonic pupil is a neurological disorder characterized by a tonically dilated pupil that reacts slowly to light but shows a more definite response to accommodation (light-near dissociation). It is frequently seen in females with absent knee or ankle jerks and impaired sweating. Other signs may include hyperopia due to accommodative paresis, photophobia and reading difficulty. Our patient presented only with a tonic pupil and no other ophthalmic complaints and hence it is ruled out.

2. Horner's syndrome results from an interruption of the sympathetic nerve supply to the eye and is characterized by the classic triad of miosis (constricted pupil), partial ptosis and loss of hemifacial sweating (anhidrosis). Our patient had features of mydriasis and absence of ptosis.



Figure 1: Right side: Hyperhidrosis Left side: Anhidrosis



Figure 2: Hyperhidrosis with hair present over right side mandibular area

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Figure 3: Anhidrosis with minimal hair over left side mandibular area



Figure 4: Left side hyperhidrotic patches

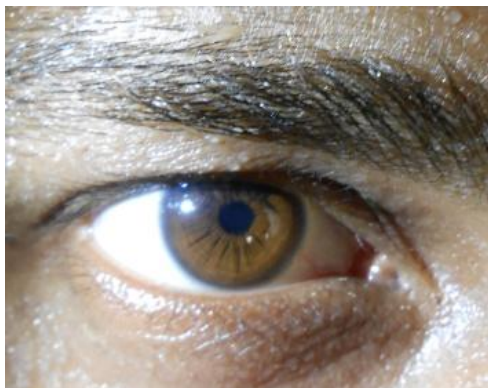


Figure 5: Right side normal pupil



Figure 6: Left side mydriatic pupil



Figure 7: Adie's tonic pupil – left eye

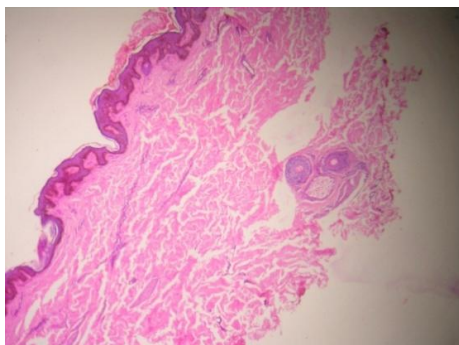


Figure 8: Presence of sweat glands-right side hyperhidrotic patch.

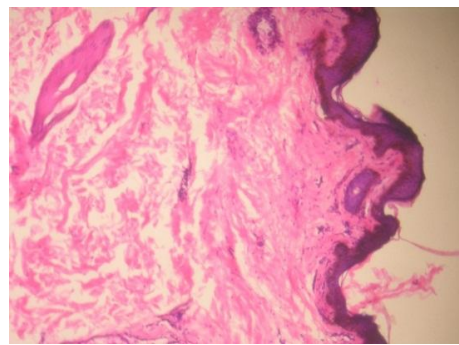


Figure 9: Absence of sweat glands - left side anhidrotic patch

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3. Hereditary sensory and autonomic neuropathy type 4 and 5 respectively are also known as congenital insensitivity to pain with anhidrosis (CIPA) and congenital insensitivity to pain with partial anhidrosis were ruled out with the presence of normal CNS functions in our patient.

Our patient had typical areas of anhidrosis and compensatory hyperhidrosis with absent tendon reflexes and tonic pupil. There were also two unusual features in our patient (i) hyperpigmentation corresponding to hyperhidrotic areas (ii) sparse hairs over left side of the beard and moustache area. Signs of autonomic dysfunction such as orthostatic hypotension, palpitation of the heart, vasovagal syncope, dyspnoea, headache, reflux esophagitis, irritable colon and psychiatric disorders may be associated with the triad of symptoms. Our patient did not have any of these associations. But owing to the complexity of the syndrome, a long follow-up of these patients is necessary. Patient was treated symptomatically with emollients and showed some improvement of xerosis after 3 months. Aluminium chloride gel (20%) also given for hyperhidrosis. Scanning of the available literature shows these unusual features present in our patient as the second patient reported in the literature. Agarwal *et al.*, (1997) have reported two cases of Ross syndrome. Sawhney *et al.*, (2004) have reported a case of variant of Ross syndrome. To the best of our knowledge, this is the second classical case of Ross syndrome with unusual features to be reported from India where a similar case was first reported by Feroze Kaliyadan *et al.*, (2009).

Conclusion

To conclude, dermatologists must be aware of Ross syndrome, which may present as its main objective symptoms with segmental hyperhidrosis and disabling heat intolerance.

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