

A Rare Case of Ross Syndrome

Sir,

A 32-year-old female presented to us with complaint of loss of sweating on most of the left side of the body along with generalized burning sensation on sun exposure. There was no history of headache, dyspnoea, palpitation, giddiness, hypotension, pain abdomen, or diarrhoea. There was no history of any trauma to the spine and syncopal attacks. Family history was not contributory.

Cutaneous examination showed anhidrosis over left side of the trunk, bilateral upper limbs, lower limbs, right side of face, palms, and soles associated with compensatory hyperhidrosis on right side of the abdomen [Figure 1] (dermatomal distribution T8–T12) and left side of face. Moreover, there was hypoplasia of bilateral 4th and 5th toe. On nervous system examination, deep tendon reflexes including knee, ankle, biceps, and supinator of both sides were absent. Blood pressure was normal in supine as well as sitting position. Rest of the cutaneous and systemic examination was normal.

Pupillary examination showed anisocoria, with the left pupil larger than the right [Figure 2]. Left-sided pupil was dilated and sluggishly reacting to light [Figure 3]. Consensual light reaction of the right pupil was normal and absent in the left pupil. Bilateral visual acuity, color vision, accommodation reflex, external ocular movements, fundus examination, and visual fields were normal. There was no ptosis. Findings of eye examination suggest Adie's pupil in the left eye.

Laboratory investigations including hemogram, liver and kidney function tests, urine analysis and thyroid function tests, HIV, VDRL, ANA, and rheumatoid factor revealed no abnormality. X-ray of chest and lumbar spine and MRI of spine was normal. Cerebrospinal fluid examination did not reveal any evidence of infection. Histology of the anhidrotic skin showed sparse sweat glands and of the hyperhidrotic region showed increased numbers of sweat glands [Figure 4].

Based on the above findings, diagnosis of Ross syndrome was made. Ross syndrome, first described by Ross in 1958, is an uncommon disorder of thermoregulation characterized by the triad of hyporeflexia or areflexia, tonic pupil, and segmental anhidrosis with or without compensatory hyperhidrosis.^[1] The first two components define the Holmes–Adie syndrome and the last component defines the Harlequin syndrome.^[2] Tonic pupil and reduced sweating can be attributed to affection of postganglionic cholinergic fibres projecting to the iris and sweat glands.^[3] The number of sweat glands may be decreased in the anhidrotic skin.^[4] Depression of deep tendon reflexes is due to dorsal root ganglionic degeneration and spinal interneuron loss. Anhidrosis may be accompanied by other alterations of the autonomic nervous system such as cardiac dysautonomia, diarrhea, and coughing.^[5] Ross syndrome may be associated with cytomegalovirus infection, in which the symptoms improved partially with the infection resolution.^[6]

Our patient presented with all typical features of Ross syndrome along with an additional finding of toe



Figure 1: Anhidrosis on the left side with compensatory hyperhidrosis on the right side (T8–T12)



Figure 2: Sluggishly reacting left pupil and normally reacting right pupil to light



Figure 3: Closer view of the dilated left tonic pupil

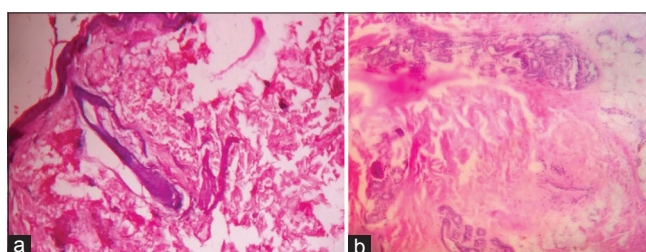


Figure 4: (a) Anhidrotic skin showing absent eccrine glands; (b) Hyperhidrotic skin showing abundant eccrine glands; (H and E, ×100)

Table 1: Differential diagnosis of Ross syndrome			
Signs and symptoms	Holmes-Adie syndrome	Horner syndrome	Hereditary sensory autonomic neuropathy
Anhidrosis	Present, impaired sweating	Present, hemifacial sweating	Present
Ophthalmic examination	Adies tonic pupil, photophobia, reading difficulty, accommodation paresis	Miosis, ptosis	Normal
Deep tendon reflex and other nervous system abnormality	Absent knee and ankle jerk, reading difficulty	Normal	Congenital insensitivity to pain

hypoplasia. The differential diagnoses of anhidrosis such as Shy Drager syndrome, multiple sclerosis, harlequin syndrome, diabetes mellitus, leprosy, and polyneuropathies were excluded as tonic pupil and areflexia are not features of these disorders. We also considered and ruled out the differentials of Holmes–Adie syndrome, Horner’s

syndrome, and hereditary sensory autonomic neuropathy [Table 1]. The case is reported due to its rarity.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Manoj K. Sharma, Savera Gupta, Sumit Yadav, Ramesh Kumar

Department of Dermatology, Venereology, Leprology, Government Medical College, Kota, Rajasthan, India


Address for correspondence:

Dr. Ramesh Kumar,
Department of Dermatology, Venereology and Leprology,
Government Medical College Kota,
Rajasthan - 324 005, India.
E-mail: dr.manojsharmak@gmail.com

References

1. Shin RK, Galetta SL, Ting TY, Armstrong K, Bird SJ. Ross syndrome plus: Beyond horner, Holmes-Adie, and harlequin. *Neurology* 2000;26:1841-6.
2. Nolano M, Provitera V, Perretti A, Stancanelli A, Saltalamacchia AM, Donadio V, *et al.* Ross syndrome: A rare or a misknown disorder of thermoregulation? A skin innervation study on 12 subjects. *Brain* 2006;129:2119-3.
3. Weller M, Wilhelm H, Sommer N, Dichgans J, Wiethölter H. Tonic pupil, areflexia, and segmental anhidrosis: Two additional cases of Ross syndrome and review of the literature. *J Neurol* 1992;239:1432-59.
4. Metta AK, Athanikar SB, Ramchandra S, Mohammad S. Ross syndrome. *Indian J Dermatol Venereol Leprol* 2009;75:414-6.
5. Chemmanam T, Pandian JD, Kadyan RS, Bhatti SM. Anhidrosis: A clue to an underlying autonomic disorder. *J Clin Neurosci* 2007;14:94-6.
6. Nagane Y, Utsugisawa K. Ross syndrome associates with cytomegalovirus infection. *Muscle Nerve* 2008;38:924-6.

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

Access this article online	
Website: www.idoj.in	Quick Response Code 
DOI: 10.4103/idoj.IDOJ_356_16	

How to cite this article: Sharma MK, Gupta S, Yadav S, Kumar R. A rare case of ross syndrome. *Indian Dermatol Online J* 2017;8:370-2.

Received: October, 2016. **Accepted:** February, 2017.

© 2017 Indian Dermatology Online Journal | Published by Wolters Kluwer - Medknow