(International Print/Online Journal)

SJIF IMPACT FACTOR: 5.565
PUBMED-National Library of Medicine ID-101739732

ISSN (Print): 2209-2870
ISSN (Online): 2209-2862

International Journal of Medical Science and Current Research (IJMSCR)
Available online at: www.ijmscr.com
Volume 5, Issue 5 , Page No: 620-623
September-October 2022

IJMSCR

A Rare Case Of Ross Syndrome - A Report

[1]Dr. Divya Mani , [2]Dr. Damayandhi Kaliyapermal, [3]Dr. Rajkiran Takharya, [4]Dr. Jude Ernest Dileep,
[1]Postgraduate, [2]Associate Professor, [3]Senior Resident, [4]Assistant Professor,
Department of Dermatology,
[1,2,4]Aarupadai Veedu Medical College And Hospital, Pondicherry.
[3]Manipal Tata Medical College, Jamshedpur.

***Corresponding Author:**
Dr. Jude Ernest Dileep
Assistant Professor,Department of Dermatology, Aarupadai Veedu Medical College And Hospital, Pondicherry

Type of Publication: Case Report
Conflicts of Interest: Nil

Abstract

Introduction: Ross syndrome (RS) is a rare disorder of the peripheral autonomic nervous system that is defined by the triad: tonic pupils, reduced or loss of deep tendon reflexes (hyporeflexia or areflexia), and anhidrosis or hypohidrosis.

Case Report: A 23-year-old adult male presented to our hospital with a history of decreased sweating over the right side of his face, neck and upper trunk for the past 7 years. He also complained of increased sweating on the left side of his face and trunk along with darkening of the skin. No history of visual disturbances, chronic illness including diabetes or trauma. The patient was advised skin biopsy from both sides and biopsy was taken on both sides from the infra- scapular region. The histopathological examination showed an absence of eccrine glands on the right (anhidrotic) side whereas the presence of normal eccrine glands on the left (hyperhidrotic) side was suggestive of Ross syndrome.

Discussion: RS is a progressive autonomic disorder that can manifest in any age, ethnicity, or gender. The average mean age is 36 years and slightly predominant in women. The exact pathogenesis of RS is largely unknown. But, many think there is a possibility of nonspecific degeneration of parasympathetic neuronal structures. Anhidrosis or hypohidrosis likely stems from the degeneration of sympathetic ganglion cells or postganglionic projections. There also appears to be a loss of regulation of skin blood flow resulting in large and dilated blood vessels in the upper dermis.

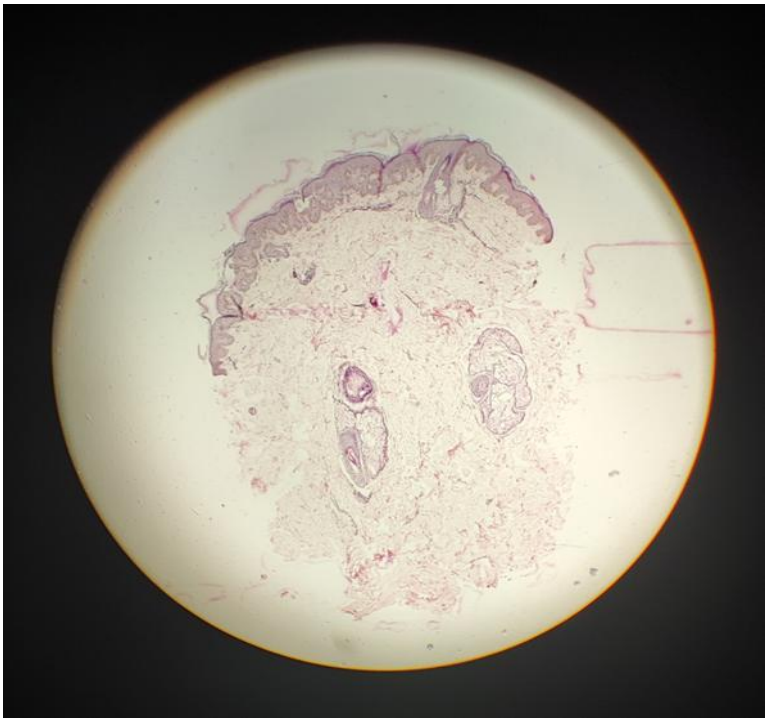
Keywords: Ross syndrome- hyporeflexia- anhidrosis

Introduction

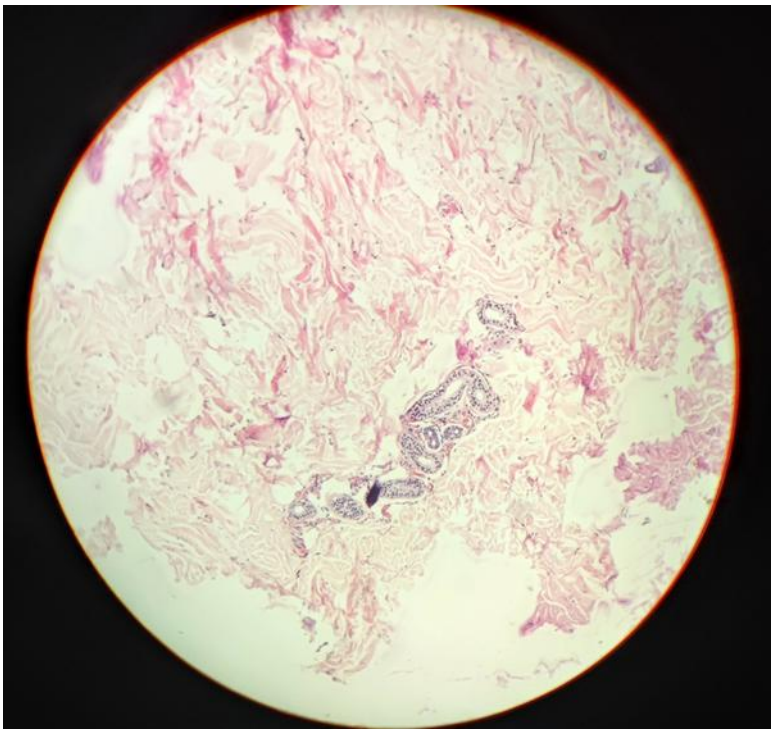
A 23-year-old adult male presented to our hospital with a history of decreased sweating over the right side of his face, neck and upper trunk for the past 7 years. He also complained of increased sweating on the left side of his face and trunk along with darkening of the skin. No history of visual disturbances, chronic illness including diabetes or trauma. On exertion, there was a lack of sweating on the right half of the face and upper trunk whereas, hyperhidrosis was noted on the left side. There was well-defined hyperpigmentation over the left side of his face, neck and upper trunk (Fig 1). Hypopigmentation and hypotrichia were noted on the right chest. Deep tendon reflexes were sluggish on both sides. On ophthalmic examination, there was a bilateral tonic pupil characterised by a sluggish reaction to light reflexes, whereas briskly reacting to accommodation reflex. Another ophthalmic examination and other system examinations were normal. All basic blood investigations were within normal limits. The patient was advised skin biopsy from both sides and biopsy was taken on both sides from the infra-scapular region. The histopathological

examination showed an absence of eccrine glands on the right (anhidrotic) side whereas the presence of normal eccrine glands on the left (hyperhidrotic) side was suggestive of Ross syndrome. The patient was treated with aluminium hydroxide to prevent excessive sweating and advised cooling measures like loose clothing, and avoiding hot environments.

Pic 1: showing absence of sweat glands with perivascular inflammatory infiltrate



Pic 2: showing presence of sweat glands



Discussion:

Volume 5, Issue 5; September-October 2022; Page No 620-623

RS is a progressive autonomic disorder that can manifest in any age, ethnicity, or gender. The average mean age is 36 years and slightly predominant in women.[2] The exact pathogenesis of RS is largely unknown. But, many think there is a possibility of nonspecific degeneration of parasympathetic neuronal structures.[3] Anhidrosis or hypohidrosis likely stems from the degeneration of sympathetic ganglion cells or postganglionic projections. There also appears to be a loss of regulation of skin blood flow resulting in large and dilated blood vessels in the upper dermis. The tonic pupil stems from damage to the ciliary ganglion or postganglionic parasympathetic nerve fibres. The deep tendon reflexes' loss may may be because of the damage to the dorsal root ganglia or due to spinal interneuron loss. Recently an autoimmune pathogenic mechanism for RS has also been proposed.[4]. A positive Serum antinuclear antibody (ANA) has been reported in a subset of RS. Another study of the opinion that RS may be an α-synucleinopathy after detecting the increased accumulation of α-Syn in autonomic nerve endings along the lesser curvature of the stomach.[5]

The diagnosis of RS is purely based on clinical presentation but the classic RS triad may not be seen initially in all patients. The tonic pupil produces variable pupillary asymmetry which is worse in the light than in the dark. The pupillary reaction to light is typically impaired but preserved to an accommodative stimulus (i.e. light-near dissociation). Pupil abnormality is typically seen on both sides in RS. The light near dissociation of the pupils is not only present in the RS. It is seen in patients with bilateral anterior afferent visual pathway disease, Argyll Robertson pupil, diabetes, or dorsal midbrain lesions.[6] The denervated sphincter muscles of the eye can show increased sensitivity to diluted pilocarpine (0.125%) . This is due to the up-regulation of receptors in a majority of the cases. Apart from the classic triad of RS , the patients will have other symptoms of autonomic dysfunction like gastrointestinal paresis, , orthostatic hypotension, Parkinsonism, impotence and sexual dysfunction and urinary incontinence .

Currently, treatment for RS is restricted mainly to symptomatic management. Treatment options for RS are similar to other primary or secondary hyperhidrosis. General cooling measures like wearing loose clothing and avoiding exposure to hot environments and avoiding heavy exercises are recommended. Aluminium chloride 10–25% is used to as an antiperspirants to prevent excess sweating but sometime it may worsen overheating.

There has been a research that also shows anti-cholinergic medications including oxybutynin and glycopyrrolate inhibit the effect of acetylcholine on the sweat glands. [7] They can be useful in reducing the sweat but on the other hand there are side effects like dry mouth, constipation, blurred vision and urinary retention. Other modalities like Botulinum toxin injections, iontophoresis, and sympathectomy have also been found to be useful in the treatment of severe hyperhidrosis. For patients with ANA positivity, Vasudevan et al presented a case of ANA positivity who was successfully treated with intravenous immunoglobulin.[4]

Conclusion:

Ross Syndrome is a rare disorder and so far only 50 cases have been reported so far in the literature. The diagnosis is done clinically and treatment is mainly symptomatic. Our case has been presented because of its rarity.

References:

1. Filikci Z, Horsten H-H, Lindelof M. Ross Syndrome: A Patient with a 23-Year History. Case Reports in Neurology. 2020;12(1). doi:10.1159/000507186
2. Nolano M. Ross syndrome: a rare or a misknown disorder of thermoregulation? A skin innervation study on 12 subjects. Brain. 2006;129(8). doi:10.1093/brain/awl175
3. Mullaaziz D, Kaptanoglu AF, Eker A. Hypohidrosis or hyperhidrosis? Ross syndrome. Dermatologica Sinica. 2016;34(3). doi:10.1016/j.dsi.2015.10.004
4. Vasudevan B, Sawhney M, Vishal S. Ross syndrome with ANA positivity: A clue to possible autoimmune origin and treatment with intravenous immunoglobulin. Indian Journal of Dermatology. 2010;55(3). doi:10.4103/0019-5154.70694
5. Ma M, Yao J, Chen Y, et al. Is Ross Syndrome a New Type of Synucleinopathy? A Brief Research Report. Frontiers in Neuroscience. 2020;14. doi:10.3389/fnins.2020.00635

Volume 5, Issue 5; September-October 2022; Page No 620-623

6. Leavitt JA, Wayman LL, Hodge DO, Brubaker RF. Pupillary response to four concentrations of pilocarpine in normal subjects: application to testing for adie tonic pupil. American Journal of Ophthalmology. 2002;133(3). doi:10.1016/S0002-9394(01)01420-9
7. Panda S, Verma D, Budania A, Bharti J, Sharma R. Clinical and laboratory correlates of selective autonomic dysfunction due to Ross syndrome. Journal of Family Medicine and Primary Care. 2019;8(4). doi:10.4103/jfmpc.jfmpc_151_19
8. The Academy uses cookies to analyze performance and provide relevant personalized content to users of our website.Categories: ArticlesNeuro-ophthalmology/Orbit.

Volume 5, Issue 5; September-October 2022; Page No 620-623