

Unilateral Anhidrosis – A Rare Entity Brought Into View by Unilateral Miliaria Rubra

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I. Introduction

Unilateral anhidrosis of face and chest and absence of flushing, also called Harlequin syndrome, is a very rare condition. It mainly involves face and upper part of the chest. It occurs due to localized sympathetic dysfunction.

II. Case Report

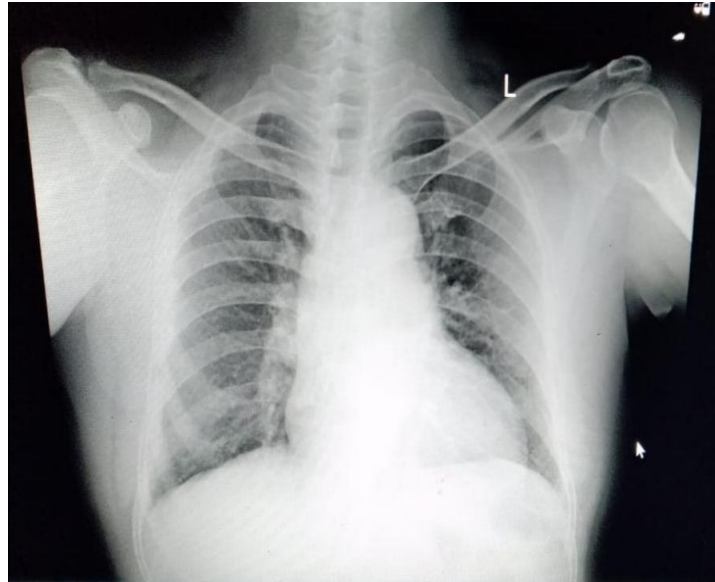
A 19 year old male patient presented with complaints of multiple small itchy lesions associated with sweating over left side of face and chest from 15 days and raised skin lesions with photosensitivity over right side of neck from 1 week. History revealed absence of sweating over right side of face and chest from 5 years of age. There was no history of abnormality in vision, weakness of limbs, sensory abnormalities, bladder dysfunction, trauma, tumor or pain in the neck region, pallor, ischemia of the fingers. On examination miliaria rubra was seen confined to left side of face and upper part of chest. Well defined hyperpigmented plaques with hypopigmented borders suggestive of polymorphic light eruption over right side of neck. Pupils, muscle power, deep tendon reflexes were normal. No sweating was seen over right side of face and chest after 20 minutes of exercise. Complete hemogram, renal function tests, liver function tests, serum electrolytes, thyroid profile were normal. Radiological investigations Chest X-ray, MRI brain and spinal cord showed no abnormalities.



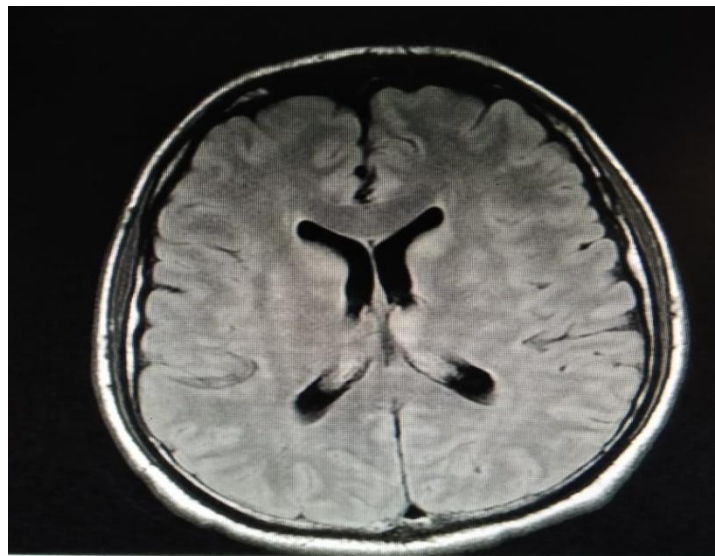
Clinical photograph showing miliaria rubra confined to left side of face



Clinical photograph showing miliaria rubra confined to left side of chest



Chest X ray revealing no abnormality



MRI brain revealing no abnormality

III. Discussion

Partial dysautonomia is characterized by abnormal regulation of the autonomic nervous system, resulting in symptoms like focal or generalized decrease in sweating, flushing, orthostatic hypotension, aberrant ocular responses. It occurs due to localized sympathetic dysfunction. It can be either primary or secondary. Such secondary causes should be excluded before diagnosing a primary condition.

PRIMARY	SECONDARY
Harlequin syndrome	Horners' syndrome
Ross syndrome	Syringomyelia
Holmes- Adies syndrome	Pancoast tumor
	Brain infarcts
	Multiple sclerosis
	Guillain – Barre syndrome
	Mediastinal tumors
	Neurogenic tumors compressing sympathetic nerve trunks
	Diabetic neuropathy
	Multiple system atrophy

Primary unilateral anhidrosis is a very rare and unique condition. Partial dysautonomias in adults are usually primary conditions where as in children are usually secondary in nature.

Harlequin syndrome is a primary condition characterized by unilateral anhidrosis of the face and upper part of chest with absence of physiological flushing (Harlequin Sign) with compensatory contralateral hyperhidrosis. Proposed level of injury is at the preganglionic sympathetic efferent fibers. Commonly presents in 2nd or 3rd decade of life with slight female preponderance. It is associated with Horner’s syndrome in 30% to 50% cases. Deep tendon reflexes will be normal. The disease usually follows a benign course.

	ROSS SYNDROME	HARLEQUIN SYNDROME	HOLMES – ADIES SYNDROME
Sex	Female = Male	Female > Male	Female > Male
Presentation	Heat intolerance	Hyperhidrosis	Photophobia, blurred vision
Cutaneous changes	Segmental anhidrosis with compensatory hyperhidrosis	Unilateral anhidrosis of face, compensatory contralateral hyperhidrosis with flushing (Harlequin sign)	Usually normal
Eye changes	Mydriasis	30 to 50% associated with Horner’s syndrome	Decreased response to light, tonic pupil
Deep tendon reflexes	Areflexia	Normal	Areflexia
Level of injury	Selective degeneration of sympathetic fibres	Preganglionic sympathetic efferents	Ciliary ganglion and dorsal root ganglion
Prognosis	Complete anhidrosis	Benign course	May progress to Ross syndrome

There is no treatment for this condition. Patients have to be counseled regarding the benign nature of the disease. If contralateral hyperhidrosis is troublesome botulinum injections can be given. Surgical management like sympathectomy, stellate ganglion block, costotransversectomy can be done in severe cases.

IV. Conclusion

Harlequin syndrome is a benign condition. It is a very rare presentation with only a few cases have been reported in the literature. Knowledge of the causes of unilateral anhidrosis or hypohidrosis is essential to rule out the secondary causes and initiation of early treatment if there is any.

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