ANHIDROSIS AS THE FIRST SIGN OF ROSS SYNDROME

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The Ross syndrome was first described in 1958[1], as a degenerative autonomic nervous system disorder defined by the triad: widespread anhidrosis, diminished deep tendon reflexes, and Adie's tonic pupils[1,2]. The most disturbing symptoms are segmental compensatory hyperhidrosis and heat intolerance. This article reports a 44-year-old male with a 19-year history of progressive anhidrosis with heat intolerance, as the mainly complaint. A segmental patch of compensatory hiperhidrosis was present in the lower abdominal T11–12 dermatomes.

The significance of our case report centers on the recognition of the skin lesions that lead to the diagnosis of an autonomic nervous disease.

CASE

A 45-year-old male presented with a 19-year history of anhidrosis, which first started on the upper limb. It had slowly developed up to the trunk, face and lower limbs, but has spared his lower abdomen, in the position of T11–12 dermatomes. In this place there was a segmental compensatory hyperhidrosis (Figure). He reported severe heat intolerance that obligated him to take frequent cold showers during the day. He complained of feet paresthesias, and impaired taste sensation for sweet. He has no history of postural hypotension, urinary or bowel symptoms, or any sexual disfunctions.

Physical examination revealed an irregular patch of hyperhidrosis, erythema and hypothermia on the lower back, in the T11–T12 dermatomes, and anhidrosis elsewhere (Figure). Neither the affected area nor another area of the skin showed signs of dystrophy or sensory impairment. The deep tendon reflexes were absent in both upper and lower limbs. Pupillary examination showed anisocoria, with the right pupil larger than the left. The right pupil reacted poorly and slowly to light, but the reaction to near sight was preserved. Once constricted, the right pupil remained with a tonic constriction, dilating slowly when the illumination was removed. The remainder of the neurologic examination was normal, including tests for postural hypotension. Nerves conduction study was normal without any evidence of peripheral neuropathy. Tests for sympathetic skin reflexes were absent to stimulation in upper and lower limbs. The Tilt test was normal. The cerebrospinal fluid analysis including VDRL test was normal. The diagnosis of Ross syndrome was made based on the clinical symptoms, neurologic signs, and complementary investigations. The patient was treated symptomatically with emollients and showed some improvement of xerosis after 3 months of treatment. Despite the skin symptom improvement, the heat intolerance obligated the patient to quit his job as a bus driver.

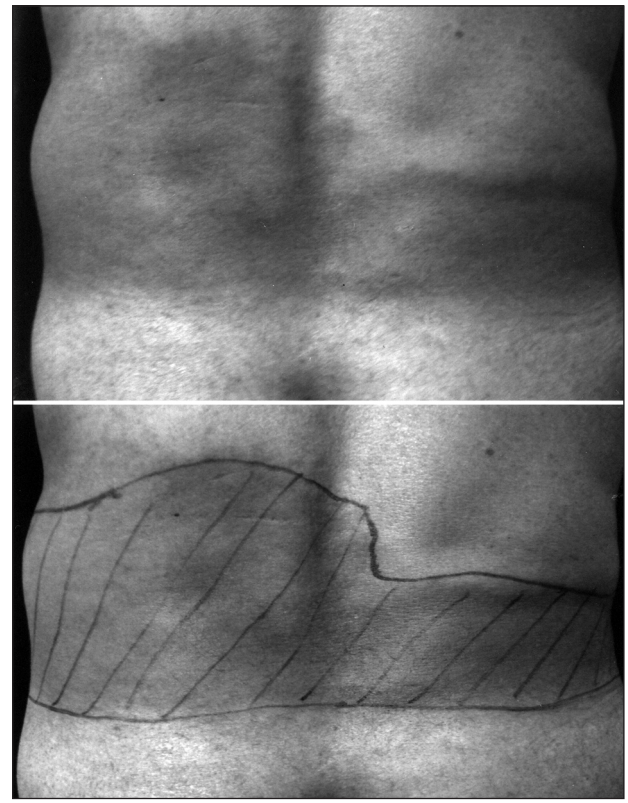

Figure. A well defined striking patch with compensatory hyperhidrosis located in lower back region.

This research project has been approved by the ethics committee of our Institute of Neurology and conforms to the pro-

ANIDROSE COMO PRIMEIRO SINAL DA SÍNDROME DE ROSS

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visions of the Declaration of Helsinki in 1995 (as revised in Edinburgh 2000).

The patient agrees with the submission of the pictures showing the skin lesion. Consent in the form of a signature was received by the authors.

DISCUSSION

Ross syndrome was first described by Ross[1], in 1958. It comprises a triad of tonic pupil, hyporreflexia and widespread anhidrosis with or without compensatory hyperhidrosis[2]. Some authors argue that the classic triad was first described ten years earlier, by Hans Hüetlin[3]. Anhidrosis may be accompanied by other alterations of the autonomic nervous system as cardiac dysautonomia[4]. Nolano et al.[2] found a complex and progressive involvement of cutaneous sensory and autonomic innervation underlying the impairment of heat production and heat dissipation. This occurs through both loss of sweating and impairment of cutaneous blood flow regulation. The pathophysiology underlying Adie's pupil is post-ganglionic denervation of cholinergic fibers between the ciliary ganglion and the iris sphincter muscle[5]. Depression of deep tendon reflexes is due to dorsal root ganglionic degeneration and spinal interneuron loss. It has been demonstrated the involvement of unmyelinated epidermal sensory fibers in patients with Ross syndrome, suggesting a degenerative and progressive disorder that involves autonomic, unmyelinated and myelinated fibers[6].

The course is usually chronic, with a possible expansion of the dyshidrotic area. Natural history is frequently slowly progressive, sometimes as long as 50 years[7]. However, Nagane et al.[8] have recently described a transitory form of Ross syndrome associated with cytomegalovirus infection, in which the symptoms improved partially with the infection resolution.

The therapeutic options included video-assisted thoracoscopic sympathectomy[9], iontophoresis[10], local instillation of botulinum toxin type A and 0,5% glycopyrrolate aquous cream[11]. The last is effective by its cholinergic receptor antagonistic effects thereby reducing sweet production rather than blocking sweat ducts, as in iontophoresis.

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REFERENCES

1. Ross AT. Progressive selective sudomotor denervation: a case with coexisting Adie's syndrome. Neurology 1958;8:809-817.
2. Nolano M, Provitera V, Perretti A, et al. Ross syndrome: a rare or a misknown disorder of thermoregulation? A skin innervation study on 12 subjects. Brain 2006;129:2119-2131
3. Weller M, Dichgans J. Erstbeschreibung des Ross-Syndroms durch Hans Hüetlin. Nervenarzt 1998:69:540.
4. Chemmanam T, Pandian JD, Kadyan RS, Bhatti SM. Anhidrosis: a clue to an underlying autonomic disorder. J Clin Neurosci 2007;14:94-96.
5. Raza N, Dar N, Mustafvi S, Zafar O. Ross syndrome with generalized anhidrosis and localized disabling compensatory hyperhidrosis. Ann Saudi Med 2008;28:53-54.
6. Perreti A, Nolano M, De Joanna G, et al. Is Ross syndrome a dysautonomic disorder only? An electrophysiologic and histologic study. Clin Neurophysiol 2003;114:7-16.
7. Beier C, Ernemann U, Gerloff C. Moving along with Ross syndrome--a patient with a 50 years history. J Neurol 2004;251: 1402-1403.
8. Nagane Y, Utsugisawa K. Ross syndrome associates with cytomegalovirus infection. Muscle Nerve 2008:38:924-926.
9. Serra MM, Callejas MA, Valls SJ, Grimalt SR, Rubio GM, Inglesias SM. Surgical treatment for compensatory hyperhidrosis in Adie syndrome. Arch Bronconeumol 2004;40:97-99.
10. Reinauer S, Schauf G, Hölzle E. Ross syndrome: treatment of segmental compensatory hyperhidrosis by a modified iontophoretic device. J Am Acad Dermatol 1993;28:308-312.
11. Bajaj V, Haniffa M, Reynolds NJ. Use of topical glycopyrrolate in Ross syndrome. J Am Acad Dermol 2006;55:111-112.