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Abstract: Ross syndrome is a rare disease characterized by peripheral nervous system dysautonomia with selective degeneration of cholinergic fibers. It is composed by the triad of unilateral or bilateral segmental anhidrosis, deep hyporeflexia and Holmes-Adie's tonic pupil. The presence of compensatory sweating is frequent, usually the symptom that most afflicts patients. The aspects of the syndrome are put to discussion due to the case of a male patient, caucasian, 47 years old, with clinical onset of 25 years.
Keywords: Hypohidrosis; Hyperhidrosis; Hereditary sensory and autonomic neuropathies; Sweating; Sweating Sickness; Tonic pupil

CASE REPORT

White, 47-year-old male patient with prior systemic hypertension, had been taking captopril as daily medication. He was referred by the Neurology department because he had presented a clinical picture of anhidrosis over the right hemiface and hemithorax for 25 years, associated with diminished visual acuity in the left eye. Ten years ago it progressed with extension of the area with anhidrosis to the left hemibody and onset of hyperchromatic macule, intense hyperhidrosis involving the lower abdomen, lumbar region, flanks and right lower limb (Figure 1). The Minor's test was carried out to show profuse sudoresis contrasting with the anhidrotic areas (presence of blue color on places where there is sudoresis – Figure 2). Examination of the pupil revealed anisocoria, with the left pupil larger than the right one. At the physical examination no sensitive or motor neurological alterations were observed. The histopathologic test did not demonstrate significant difference in the quantity of sweat glands between the anhidrotic and hyperhidrotic areas, or in the population of melanocytes and melanosomes, revealing absence of pigment leakage.

Figure 1: Hyperchromatic macule with intense hyperhidrosis affecting the lower abdomen, lumbar region and flanks

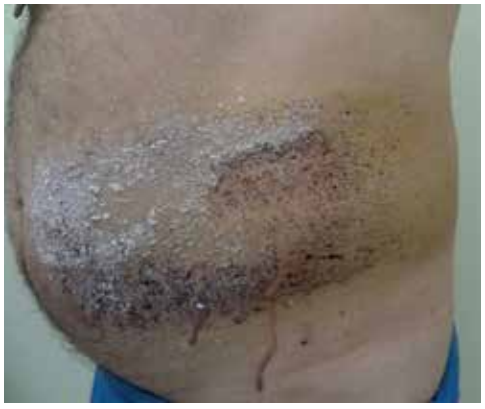

Figure 2: Minor's test: bluish staining show locations where there is sudoresis

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DISCUSSION

Ross Syndrome is a rare disease of unknown cause, first described by Ross in 1958, which represents dysautonomia of the peripheral nervous system with selective degeneration of cholinergic fibers.[1,2] It is characterized by the triad of unilateral or bilateral segmental anhidrosis, deep hyporeflexia and Holmes-Adie's tonic pupil, which is distinguished by a mydriatic pupil, unresponsive to light or accommodation. [1,3,4,5] In cases where there is extensive anhidrosis, the remaining eutrophic areas with functioning eccrine sweat glands may present compensatory hyperhidrosis, the symptom that most distresses the patients, who also become uncomfortable with heat intolerance.[4,5,6] The hyperchromia observed seems to be pigmentation similar to notalgia paresthetica, in face of the histologic findings.

There are other partial dysautonomias, represented by a spectrum of diseases with some common clinical characteristics, such as the Holmes-Adie syndrome, defined by tonic pupil, hyporeflexia or areflexia, Harlequin syndrome, characterized by hypohidrosis and facial flushing in response to heat, exertion or emotional factors, and Horner's syndrome by anhidrosis, ptosis, miosis and enophthalmus. [1,4,7]

The diagnosis of Ross syndrome is made based on clinical symptoms, neurological signs and supplementary investigation.[8]

The course of this syndrome progresses slowly, but it is benign. The complete triad usually takes years to appear.[1] This would explain, in the reported case, the presence of only two of the three alterations characteristic of the syndrome, with possible onset of deep hyporeflexia along the years. Although the thermoregulation mechanism is intensely compromised, limiting the scope of social activities of the patients, such condition does not represent risk of death, as long as the patients are oriented to develop strategies to avoid the elevation of body temperature.[8] ❑

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