Introduction

Tonic pupil syndrome, described by Adie in 1932, consists of benign ophthalmoplegia, which may present in isolation, leading to accommodation impairment, or in association with other neurological defects. Iridoplegia coexisting with hypo- or areflexia is known as Holmes-Adie syndrome, whereas the triad consisting of tonic pupil, areflexia, and segmental hypohidrosis is known as Ross syndrome. These syndromes can also be associated with other alterations of the autonomic nervous system such as orthostatic hypotension or vasovagal syncope.

Key words: Hyperhidrosis. Adie pupil. Sympathectomy.

Ross syndrome is characterized by a triad of tonic pupil, areflexia and segmental hypohidrosis. Hypohidrosis may be accompanied by contralateral hyperhidrosis, probably due to a compensatory mechanism. We report a case of Ross syndrome with socially disabling left-sided hyperhidrosis. Sympathectomy of the second and third thoracic ganglia was performed with satisfactory results. With excellent results in primary hyperhidrosis and very low morbidity, thoracic sympathectomy is the definitive treatment for hyperhidrosis.

While hypohidrosis causes little discomfort, patients frequently complains of contralateral hyperhidrosis which is probably due to a compensatory mechanism. We report the case of a man with socially incapacitating left-sided hyperhidrosis associated with Ross syndrome. The patient underwent a sympathectomy of the third thoracic ganglion with satisfactory results.

Tratamiento quirúrgico de la hiperhidrosis compensadora en el síndrome de Adie

La tríada consistente en pupila tónica de Adie, arreflexía osteotendinosa e hipohidrosis segmentaria se conoce como síndrome de Ross. La hipohidrosis puede ir acompañada de un exceso de sudación en el hemicuerpo contralateral. Esta hiperhidrosis es probablemente debida a un mecanismo de compensación. En este artículo presentamos el caso de un paciente con síndrome de Ross cuyo motivo de consulta fue hiperhidrosis socialmente incapacitante en el hemicuerpo izquierdo. El paciente fue intervenido quirúrgicamente, realizándose una simpatectomía del segundo y tercer ganglios torácicos con resultados clínicamente satisfactorios. La simpatectomía torácica es un tratamiento definitivo de la hiperhidrosis, con unos resultados excelentes en la hiperhidrosis primaria y un muy bajo índice de morbilidad.

Palabras clave: Hiperhidrosis. Pupila de Adie. Simpatectomía.

Clinical Observation

A 25-year-old male reporting no use of tobacco, alcohol, or street drugs and with an unremarkable medical history complained of craniofacial hyperhidrosis and shoulder hyperhidrosis of 6 years’ duration. His profuse sweating caused evident social distress, manifested by efforts to hide the constant wetness of his left side by wearing loose-fitting clothes.

The marked distinction between left-sided facial hyperhidrosis and right-sided hypohidrosis was immediately apparent to inspection, with vertical midline demarcation. The...
skin on the right side of the face and hand was dry whereas the left side was shiny and wet. Differences between the 2 sides were less accentuated in the lower extremities.

Physical examination showed the right pupil to be unresponsive to light. The left pupil’s response was normal. No other alterations in cranial nerve or in autonomic nerve function was observed. However, deep tendon reflexes were absent in the upper and lower extremities.

Nuclear magnetic resonance of the brain was normal thus ruling out possible vascular and compressive lesions. An electrophysiologic brainstem function test and a sweat test of sympathetic skin response were performed.

Trigeminofacial reflexes were unaltered, with normal latency and amplitude in blink and jaw reflexes (Table). However, the current perception threshold score of the right supraorbital nerve was twice that of the left (6 to 3 mA). A thermal threshold analysis using Thermotest (Somedic, Stockholm, Sweden) equipped with a 6.2 cm rectangular Peltier probe was performed to better detect right-left side differences in perception of pain caused by heat. The mean thermal threshold was significantly greater in the right side of the face compared to the left although no difference was observed in the hands (Figure 1).

The sweat test indicated that right hand response was absent to ipsi- and contralateral median nerve stimuli as well as to supraorbital nerve stimuli (Figure 2). In contrast, left hand response was normal.

The patient underwent a sympathectomy of the second and third thoracic ganglia by means of video-assisted thermocoagulation. He was discharged on day 2 post-intervention with no complications and with an evident improvement in hyperhidrosis.

**Discussion**

Adie pupil is essentially a clinical diagnosis. Pupillary hypofunction manifests as slow pupil constriction followed by gradual dilation lasting minutes or even hours. The incidence peaks between the third and fifth decades with no significant differences between the sexes. Adie pupil is unilateral in 90% of cases.

It is believed that the pathophysiology underlying Adie pupil is post-ganglionic denervation of parasympathetic cholinergic fibers between the ciliary ganglion and the iris and aberrant reinnervation of the iris through fibers destined for the ciliary muscle. Diagnosis is established upon obtaining an intense and rapid pupillary constriction by instilling a solution of 2.5% methacholine and 0.125% pilocarpine.

Ross syndrome patients can present bilateral Adie pupil in half of cases. Moreover, up to 50% of Holmes-Adie syndrome patients present asymptomatic hypohidrosis. It is therefore possible that these syndromes constitute a single entity with various types of presentation. Onset of hyporeflexia in Holmes-Adie syndrome is progressive, bilateral, and symmetric and is not accompanied by motor or sensory alterations. Although the pathophysiology of areflexia is not clear, it has been suggested that the explanation may lie in presynaptic degeneration of dorsal root fibers, which transmit afferent impulses to anterior horn cells.

Hypohidrosis in Ross syndrome patients is usually segmental with progressive onset and is probably due to the degeneration of postganglionic sympathetic fibers leading to the sweat glands. An efferent pathway alteration of the sympathetic sweat gland response, which was demonstrated in the neurophysiological test function was observed. However, deep tendon reflexes were absent in the upper and lower extremities.
of our patient, is consistent with that hypothesis. In other cases this alteration has been demonstrated by the absence of sweat gland response after subcutaneous injection of pilocarpine.\(^8\)

Compensatory hyperhidrosis in Ross syndrome has been described,\(^3,9\) though its pathophysiology is still unknown. Hyperhidrotic reactions may be observed in patients with lesions in the peripheral nervous system, such as peripheral axonopathies,\(^10\) or in the central nervous system, as with Wallenberg syndrome.\(^11\) In all such cases, hyperhidrosis is interpreted as an exaggerated reaction of sweat glands that remain undamaged.

Few cases of patients treated for compensatory hyperhidrosis are described in the literature. Our patient sought treatment due to social and occupational problems deriving from hyperhidrosis. The treatment of choice was video-assisted thoracoscopic sympathectomy.\(^12\) Other therapeutic options are iontophoresis\(^13\) and local instillation of botulinum toxin type A,\(^14\) two treatments that are free of side affects, although the resulting improvement is usually partial and transitory and treatment itself is relatively uncomfortable. Iontophoresis involves the application of weekly maintenance doses while the effect of botulinum toxin usually lasts from 3 to 6 months. Thoracic sympathectomy is the definitive treatment for hyperhidrosis: results are excellent in primary hyperhidrosis\(^15\) and the morbidity rate is very low. Our patient was discharged 48 hours after intervention and returned to work within a few weeks.

REFERENCES