Pourfour du Petit Syndrome Associated With a Cervical Vertebral Anomaly

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Abstract: Pourfour du Petit syndrome is a rare dysautonomic disorder characterized by mydriasis, eyelid retraction, and hyperhidrosis and is caused by irritative stimulation of the sympathetic cervical chain. The authors describe a 45-year-old woman with iris heterochromia, who presented with episodes of ipsilateral mydriasis and hyperhidrosis and was found to have a cervical vertebral anomaly, probably present since birth, as the cause of Pourfour du Petit syndrome.

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Pourfour du Petit syndrome (PdPS) is a rare disorder characterized by mydriasis, eyelid retraction, and hyperhidrosis. These clinical findings are the opposite of those accompanying Horner syndrome (HS), and PdPS also is known as “reverse HS” (1).

Since the first description of PdPS (2), it has been thought that an irritative lesion of the cervical sympathetic chain, resulting in sympathetic hyperactivity, is responsible for the dysautonomic syndrome. Like HS, PdPS is indicative of a disorder of the oculosympathetic pathway. At times, it is thought that PdPS can precede HS (3,4), although this has yet to be reported. Case of PdPS have been associated with cervicothoracic pathology, including cervical tumors (5,6), trauma (7), and surgical procedures (8,9). Our patient is unusual as none of these predisposing factors were present.

CASE REPORT

A 45 year-old woman reported multiple episodes over 10 years of right pupillary mydriasis and blurred vision associated with ipsilateral hemifacial hyperhidrosis, triggered by exercise and stressful situations. There was no significant medical or surgical history nor a history of trauma.

On neuro-ophthalmic examination, pupils were 3.5 mm, right eye, and 2.5 mm, left eye, without ptosis or lid retraction. Direct and consensual pupillary light responses were normal, and eye movements were intact. Visual acuity, intraocular pressures, and funduscopy were normal, whereas slit-lamp examination showed iris heterochromia with the right iris being more pigmented than the left. The right pupil showed no supersensory to 0.125% pilocarpine drops, and the responses to 1% pilocarpine and 4% cocaine drops were normal and bilaterally symmetric.

Within minutes of the patient starting physical exercise (the patient ascended and descended the stairs up to the sixth floor twice), we found that the right pupil dilated to 6.5 mm with ipsilateral hemifacial hyperhidrosis (Figs. 1, 2).

Laboratory data, including thyroid function tests, chest radiography, and brain computed tomography were within the normal limits. Magnetic resonance imaging (MRI) of the spine showed narrowing of both the C6–C7 disc space and the right C6–C7 neuroforamen (Fig 3). Because the patient’s clinical symptoms were not disabling and episodes were triggered only by exercise, no treatment was given. The patient remained stable over 6 months of follow-up.

DISCUSSION

At the beginning of the 18th century, François Pourfour du Petit, a French military surgeon, examined many soldiers with wartime neck injuries. He noted signs of increased facial sympathetic activity and related these to injuries of the cervical sympathetic chain (2).

The anatomy and physiology of the cervical sympathetic pathway is well known. It is important in the regulation of...
facial temperature and sweating and in the control of the pupil size. The most common disorder of the cervical sympathetics is HS, which is caused by sympathetic paralysis and resultant pupillary miosis, ptosis, and anhidrosis (10). PdPS is an uncommon cause of unilateral mydriasis, lid retraction, and hyperhidrosis caused by hyperactivity of the ipsilateral oculosympathetic pathway. PdPS has been documented in association with intracranial aneurysms (11), nonpenetrating injuries of the cervical sympathetic chain and brachial plexus (7), interrupted aortic arch (4), cervical and thoracic tumors (5,6), maxillofacial surgery (8,9), and regional anesthetic procedures (10). To our knowledge, the association of cervical disc abnormality and PdPS previously has not been reported.

In our patient, MRI showed disc space narrowing at the C6–C7 level with right lateroforaminal narrowing and compression of the C7 nerve root. We speculate that this injury to the cervical sympathetic chain produced an irritative stimulus that resulted in oculosympathetic hyperactivity.

Our patient had iris heterochromia. Congenital HS is classically associated with iris heterochromia with the ipsilateral iris being less pigmented. This is thought to be due to attenuated iris melanocyte development from reduced noradrenaline release. In contrast, in our case, the iris on the affected side was more pigmented than the fellow eye. Congenital PdPS could produce sympathetic overactivity from birth, leading to amplified noradrenaline release, with increased melanocyte development and iris hyperpigmentation.

REFERENCES