

consistent with findings as Hylenex irrigation into the lacrimal system (as opposed to within periorbital tissues) was curative.

Although firm recommendations cannot be made on the basis of a single report, off-label irrigation of hyaluronidase may be considered in the extremely rare event of NLDO following tear trough HA injection.

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Pourfour du Petit Syndrome: A Rare Association With Cluster Headache

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Abstract: Pourfour du Petit syndrome is an uncommon cause of eyelid retraction, associated with unilateral mydriasis and hemifacial hyperhidrosis. This syndrome is caused by hyperactivity of the ipsilateral oculosympathetic pathway and needs to be recognized because it has an opposite clinical presentation but the same topographic and diagnostic value as Horner syndrome. The authors report a rare case of Pourfour du Petit syndrome associated with cluster headache and discuss pathophysiological hypotheses, clinical presentation, complementary exams, pharmacologic testing, treatment options, and prognosis. Early detection of these symptoms may lead to swift diagnosis and treatment.

Pourfour du Petit (PDP) syndrome is a rare and underdiagnosed cause of superior eyelid retraction which is necessary to recognize because it shares the same etiologies as Horner syndrome. Here, the authors describe the first case of PDP syndrome associated with cluster headache and discuss clinical features, management, and outcome of PDP syndrome. This report is compliant with the Declaration of Helsinki.

CASE PRESENTATION

A 48-year-old woman reported a 3-year history of left anisocoria. Her medical history included a 30-year history of left cluster headache treated by Sumatriptan injection. Medical

examination confirmed a PDP syndrome, with left mydriasis and upper eyelid retraction only (Fig. 1). Direct and consensual light responses were normal. The ophthalmic symptoms increased during facial paroxysmic pain and persisted all day long. No cold sensation or hemifacial pallor was reported. Visual acuity, intraocular pressures, and fundoscopy were normal. The left pupil showed no supersensitization to 0.125% pilocarpine drops and a normal response to 1% pilocarpine. Repeated thyroid tests (Table), the brain and cervical MRI scan, and the Doppler of the supra-aortic vessels were also normal.

DISCUSSION

Pourfour du Petit syndrome was first described by Francois Pourfour Du Petit, a French surgeon of the armies of Louis XIV. Soldiers brought into the military hospitals after neck injuries showed signs of increased facial sympathetic activity in the eyes and upper extremity. In PDP syndrome, the clinical findings are the opposite of those accompanying Horner syndrome: unilateral mydriasis, widening of the palpebral fissure secondary to superior eyelid retraction, proptosis, pale sclera, pale and cool facial skin with increased sweating. However, the symptoms are often uncompleted. An association with iris hyperpigmentation was also reported.¹

It is thought that irritative stimuli of the sympathetic fibers are responsible for this focal dysautonomic syndrome, resulting in a sympathetic hyperactivity. Causes are also the same as in Horner's syndrome: cervical or cerebral vascular anomalies, trauma, tumors, regional surgical, or anesthetic procedures (Fig. 2).

The case illustrates a pathologic association between cluster headache and PDP syndrome, raising the question of a novel autonomic dysfunction in cluster headache. Such a clinical association has not been described previously, but about 10% of patients with cluster headache exhibit a Horner syndrome.² Pourfour du Petit syndrome was also described in association with migraine and with recurrent paroxysmal right facial pain, trigeminal neuralgia pain,³ and occipital neuralgia.⁴ Acute pain due to angle-closure glaucoma was also described secondary to PDP syndrome. In contrast to the case, the pain was not directly associated with PDP syndrome or sympathetic



FIG. 1. Photograph of the patient's face in pathological condition combining unilateral left cluster headache and Pourfour du Petit syndrome: anisocoria and left upper eyelid retraction.

Timeline of thyroid tests

Thyroid tests	Date	Values	Normal values
TSH	Baseline	1.79 mU/L	0.27–4.2 mU/L
	+4 months	1.20 mU/L	
	+9 months	1.37 mU/L	
	+18 months	2.02 mU/L	
	+24 months	1.49 mU/L	
Free thyroxine (T4)	Baseline	17.7 pmol/L	12–22 pmol/L
TSH receptor antibodies	Baseline	Negative	<1.8 U/L

TSH, thyroid stimulating hormone

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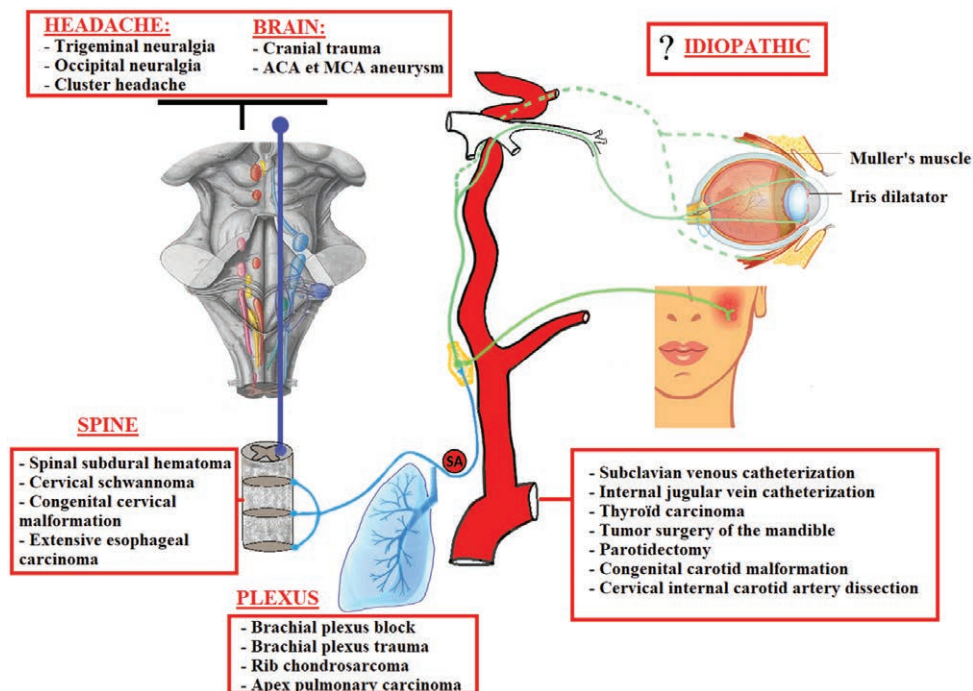


FIG. 2. Known causes of Pourfour du Petit syndrome. Modified with permission from *Headache*. 2014;54:373–7.

hyperactivity but was rather a consequence of a rare complication of this syndrome.⁵

Mydriatic pupil shows no supersensitization to 0.125% pilocarpine drops, and the response to 1% pilocarpine is bilaterally symmetric. This pharmacologic testing eliminates the diagnosis of Adie's syndrome and pharmacologically dilated pupils. Any thyroid disorder must also be excluded biologically. A single brain angio-MRI extending to the T2 level in the chest is necessary to rule out causative factors of sympathetic dysfunction.⁶

Pourfour du Petit has a variable duration, outcome is highly unpredictable, and the prognosis is not clear: physical signs may persist for an indefinite time^{7,8} or may resolve in a few months once the underlying stimulus has stopped.⁹ The treatment is primarily etiologic; myotic drops are relatively inefficient, upper thoracic sympathectomy¹⁰ and stellate ganglion block¹¹ could be considered only in severe cases. Unusual presentations with major eyelid retraction rarely require a tarsorrhaphy, pomades, and nocturnal eyelid occlusion.¹² It seems reasonable to assume that a mullerectomy should be performed in persistent eyelid retraction but was never realized in the literature. Pourfour du Petit syndrome is a very particular and rare cause of superior eyelid retraction, often transient and under diagnosed. Early detection of these symptoms may lead to swift diagnosis and treatment.

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Complete Excision of a Simple Dacryops Using Fibrin Sealant and Trypan Blue Mixture

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