

LETTER TO THE EDITOR

Pourfour du Petit syndrome after medullary thyroid carcinoma surgery[☆]



Síndrome de Pourfour du Petit tras cirugía de carcinoma medular de tiroides

Pourfour du Petit syndrome (PdP) or oculosympathetic spasm consists of unilateral mydriasis, upper eyelid retraction and hyperhidrosis related to irritation and hyperstimulation of the oculosympathetic pathway in the neck. It is important to recognise PdP, as it has the same topographic and diagnostic value as its opposite clinical presentation, Horner's syndrome, caused by a lesion resulting in a deficiency of this same pathway.

It was first reported by Italian physicist Serafino Biffi in 1846.¹ Even then, he identified irritative stimulation of the sympathetic nerve fibres in the neck adjacent to the carotid wall as responsible for the syndrome. However, it owes its name to Francois Pourfour du Petit (1664–1741), a French military surgeon who studied the sympathetic pathway in the neck and its implications for the eye in the early 18th century.

Knowledge of PDP has a dual importance. On the one hand, it must be recognised in order to prevent the common error of mistaking the pathological finding for miosis and ptosis on the contralateral side, in the form of a false Horner's syndrome on the healthy side. On the other hand, it may be the only clinical indicator of potentially serious disorders such as carotid dissection. In that regard, although all diseases and conditions that cause Horner's syndrome could potentially present as PdP, very few cases of PdP have been reported in the literature and those that have been reported are heterogeneous. They may be iatrogenic, after anaesthetic procedures² or surgical techniques in that area,^{3,4} or they may be secondary to a range of disorders (carotid dissection,⁵ phenomena of compression,^{6,7} or headache⁸).

We report the case of a 31-year-old woman recently diagnosed with multiple endocrine neoplasia, type 2A (MEN 2A),

a syndrome involving C634S mutation in the RET oncogene. She underwent surgery for a right pheochromocytoma and further surgery for medullary thyroid carcinoma with cervical lymph node metastases. The second procedure consisted of a total thyroidectomy plus central and bilateral functional lymphadenectomy.

On day five after thyroid surgery, the patient was found to have anisocoria with right reactive mydriasis with ipsilateral eyelid retraction, which she had not had previously (Fig. 1A). The rest of the examination was strictly normal.

A duplex study of the supra-aortic trunks showed no morphological or haemodynamic abnormalities of the right carotid axis from its origin in the brachiocephalic trunk to the mandibular angle. Magnetic resonance imaging of the neck showed no abnormalities of the morphology or lumen of the common carotid artery, internal carotid artery or vertebral artery and no signs suggestive of intimal or sub-adventitial dissection.

In view of the above, the patient was diagnosed with Pourfour du Petit syndrome thought to be secondary to surgical manipulation, owing to a mechanism of elongation without rupture of the arterial tissue. On the one hand, this diagnosis was made due to the absence of signs of arterial damage in the imaging tests. On the other hand, the clinical recovery indicated that the mechanism of irritation of the nerve fibres in the arterial adventitia was also distension, with no rupture present. In this context, PdP syndrome can also be due to other more serious processes, so the patient should be promptly examined with the appropriate imaging tests. It may be due to extrinsic compression of the artery (e.g. bruising) or to more severe trauma resulting in carotid dissection. The means that the prognosis for PdP syndrome is variable, mainly depending on the root cause. Symptoms are often transient and resolve within a few months, once the trigger stimulus has ceased; however, in some cases, they remain stable and persist over time. In any event, in view of the minimal or non-existent discomfort experienced by patients, no specific treatment is usually given.

In the case of our patient, complete resolution of the condition occurred two months after its onset. Examination in follow-up at the endocrinology clinic revealed that her pupils were equal and normally reactive to light (Fig. 1B). The patient is presently stable and asymptomatic.

[☆] Please cite this article as: González Villar E, Jiménez CM, Olmos Alemán M, Ayo-Martín Ó. Síndrome de Pourfour du Petit tras cirugía de carcinoma medular de tiroides. *Endocrinol Diabetes Nutr.* 2021;68:366–367.



Figure 1 (A) Day five after thyroid surgery, with right mydriasis and ipsilateral upper eyelid retraction. (B) Follow-up two months after thyroid surgery, with complete resolution of the condition.

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