

Case report

Unilateral hyperhydrosis in Pourfour du Petit syndrome

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Abstract

Upper limb hyperhydrosis is an idiopathic disease with bilateral involvement. However, Pourfour du Petit syndrome, the opposite of Horner syndrome, may result in unilateral upper limb hyperhydrosis. It occurs following hyperactivity of the sympathetic cervical chain as a consequence of irritation secondary to trauma. We report herein two cases with Pourfour du Petit syndrome showing unilateral upper limb hyperhydrosis. The patients presented with right-sided mydriasis and ipsilateral hemifacial hyperhydrosis. The onset of disease was followed by a trauma in both patients. They underwent upper thoracic sympathectomy with favorable outcome. A history of an antecedent trauma in patients with unilateral upper limb hyperhydrosis and anisocoria may imply a possible diagnosis of Pourfour du Petit syndrome.

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Keywords: Pourfour du Petit syndrome; Upper limb hyperhydrosis; Sympathectomy**1. Introduction**

Upper limb hyperhydrosis is a common functional disorder, which may pose serious psychologic, social, and occupational problems. It occurs as bilateral involvement of the upper extremities. Although conservative treatment modalities offer only minimal and temporary relief of symptoms, thoracic sympathectomy, particularly a videothoroscopic procedure appears to be the optimal treatment of choice with a success rate ranging from 94 to 98% in the management of upper limb hyperhydrosis [1–3].

Upper limb hyperhydrosis is almost always essential and the etiology remains unclear. The only findings regarding the etiology are that it is mostly prevalent in young adults with a slight female predilection, and a family history ranging from 12.5 to 56.5% [1]. Pourfour du Petit syndrome (PPS), the opposite of Horner syndrome (HS), is often initiated by a stimulating lesion of cervical sympathetic chain which may result in increased sympathetic activity, and thereby upper limb hyperhydrosis.

Although many reports appeared previously regarding upper limb hyperhydrosis, we are not aware of any report of

unilateral upper limb hyperhydrosis resulting from PPS. We herein report two cases with PPS showing upper limb hyperhydrosis. Both patients had an onset of disease followed by a history of trauma. They showed right-sided mydriasis and hemifacial hyperhydrosis. We tried to emphasize this exceptional syndrome, which may easily be confused with HS, as a causative factor in the etiology of upper limb hyperhydrosis.

2. Case reports**2.1. Case 1**

A 49-year-old woman was referred with presumptive diagnosis of HS on the left side. She had anisocoria (left < right) (Fig. 1A), however she had neither ptosis nor enophthalmus on the left side to prove the possible diagnosis of HS. Her anisocoria did not change during either light or darkness. Her left pupilla did not show supersensitivity reaction to either 1/1000 aqueous epinephrine or to 2.5% phenylephrine. No difference was observed between the eyes with respect to accommodation, and the intraocular pressure was 12 mmHg in both eyes. She showed mydriasis on the right side; however she had no retraction on either eyelid.

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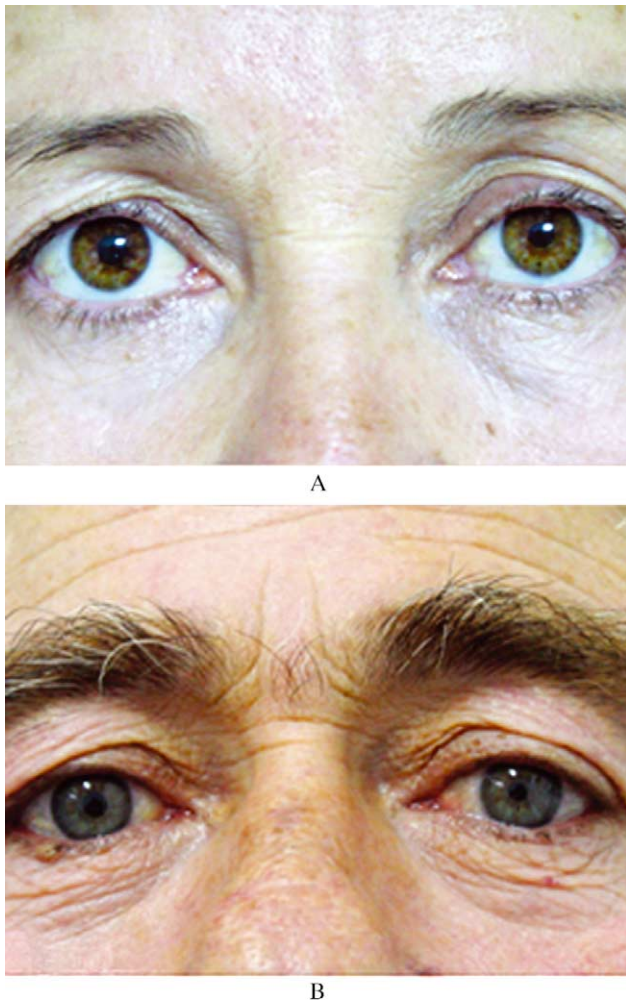


Fig. 1. (A) and (B) Anisocoria showing right-sided mydriasis in case 1 and in case 2.

She also had right upper limb hyperhydrosis (Fig. 2A) with ipsilateral hemifacial hyperhydrosis. The onset of her symptoms was 3 years before admittance following a traffic accident which she had not suffered from a major injury. In addition, she suffered from chronic headache, and tremor in her left arm, however neurological examination and cranial magnetic resonance imaging did not reveal any abnormality. Her chest X-ray and computerized tomography (CT) were normal. Laboratory data including thyroid function tests were within the normal limits. The patient underwent a videothoracoscopic right upper thoracic sympathectomy including T2–T4 ganglia. Her symptoms improved (Fig. 2B), however she showed compensatory sweating on the contralateral upper limb, which she could tolerate and vanished with time. She remains well and symptom-free at 17 months follow-up.

2.2. Case 2

A 56-year-old man was admitted with anisocoria and right-sided upper limb hyperhydrosis. He had a history of

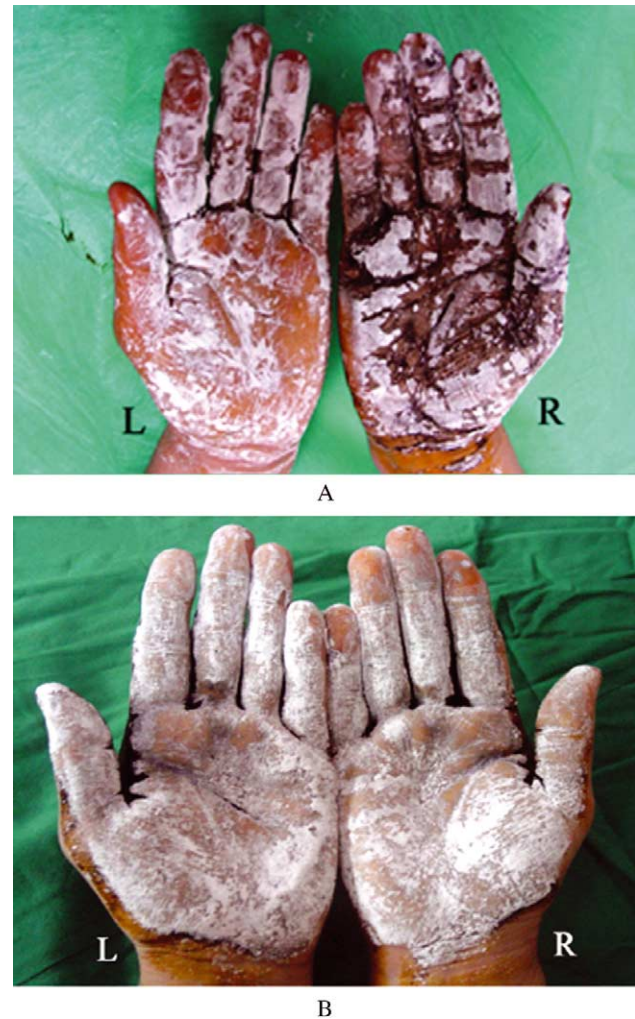


Fig. 2. (A) Preoperative iodine starch test showing hyperhydrosis of the right hand in case 1. (B) Postoperative iodine starch test showing improvement of hyperhydrosis of the right hand in case 1.

traffic accident four years before his admittance, which resulted in multiple rib fractures with bilateral hemopneumothorax requiring tube thoracostomy. His complaints started following this accident. He also had a cyst located at the dome of his liver that ultrasonography and a negative hemagglutination test for hydatid disease proved to be a simple liver cyst. His physical examination was normal except anisocoria (right > left) (Fig. 1B). Ophthalmologic examination showed similar findings to that of the first case.

His chest X-ray and CT showed posterolaterally located healed rib fractures on either side. A right-sided videothoracoscopic sympathectomy was intended; however the procedure was converted to a right lateral thoracotomy because of dense pleural adhesions found on videothoracoscopy. Intraoperative examination showed a healed rib fracture with a callus formation at the level of the head of the first rib, which showed compression on the stellate ganglion. Upper thoracic sympathectomy including T2–T4 ganglia was performed. Postoperative course was uneventful. The patient is asymptomatic five months after surgery.

3. Discussion

Horner syndrome (HS) is a well known clinical picture in thoracic surgery caused by the involvement of the cervical sympathetic chain. It occurs as a result of the damage or the involvement of the stellate ganglion most often by intrathoracic malignancies, particularly a lung tumor located at the apex of the lung, so-called Pancoast tumor. The etiology of HS consists of improper intraoperative positioning, tube thoracostomy, thoracoplasty, neck surgery and neck trauma or it may occur as a complication of thoracic sympathectomy [2,4]. The main anesthetic causes for HS are interventions for regional anesthesia such as brachial plexus block, epidural anesthesia or interpleural analgesia [4]. The components of HS are myosis, ptosis, enophthalmus of the involved eye and ipsilateral hemifacial anhidrosis. The affected eye shows increased amplitude of accommodation and a supersensitivity reaction to either 1/1000 aqueous epinephrine or to 2.5% phenylephrine. The intraocular pressure on the side with HS is expected to be 5 mm or less than the pressure of the fellow eye [5]. Ophthalmologic findings in our cases showed that they did not have HS.

PPS is reciprocal of HS and it is caused by a stimulation of the ipsilateral sympathetic cervical chain. The etiology of PPS is similar to HS and PPS can precede HS [4,6]. Previous reports have also shown that PPS might also develop secondary to a rib chondrosarcoma or to an esophageal carcinoma showing mediastinal and pleurocostal extension [6,7]. Patients often have a history of an antecedent trauma before admission [8,9]. We observed that the possible irritation of the sympathetic chain was a healed rib fracture with a prominent callus just beneath the stellate ganglion resulting from the previous trauma in our second case.

Patients with PPS often present with ocular findings those opposite of HS, and it is often reported as a unilateral mydriasis as in our cases [4]. PPS may also present with eyelid retraction and exophthalmus or even with progressive hemifacial atrophy [6,8]. PPS carries a risk for conjunctivitis, keratitis and epiphora in case of major exophthalmia. In addition, most patients with PPS have neurological symptoms such as headache and tremor as in our cases [9,10]. The finding that PPS is very likely to occur following a trauma sometimes makes the videothoracoscopic

sympathectomy procedure inappropriate as in our second case, hence an open thoracic procedure becomes inevitable.

In conclusion, although the etiology of upper limb hyperhidrosis remains obscure, anisocoria with ipsilateral hemifacial hyperhidrosis and a history of an antecedent trauma may imply a possible diagnosis of PPS in patients with unilateral upper limb hyperhidrosis. Although a videothoracoscopic approach may not be feasible in some occasions, an upper thoracic sympathectomy provides a favorable outcome in patients with PPS.

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