REGIONAL ANAESTHESIA

Case Report

Coexisting harlequin and Horner syndromes after high thoracic paravertebral anaesthesia

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A patient undergoing left mastectomy and immediate latissimus dorsi breast reconstruction under combined paravertebral block and general anaesthesia developed transient, well-demarcated, right-sided hemifacial erythema and sweating, and left-sided Horner syndrome post-operatively. This ‘harlequin’ appearance occurs because of a normal or excessive vasodilatory, thermoregulatory response to heat or emotion mediated by an intact sympathetic pathway on the erythematous side, together with relative pallor of the pharmacologically blocked side.

Keywords: anaesthetic techniques, regional, paravertebral; sympathetic nervous system, ganglion block

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Harlequin syndrome consists in sudden onset of unilateral facial flushing and sweating,1 dividing the face into two regions, sharply demarcated at the midline, i.e. a plethoric and a pale hemifacial area. We describe contralateral harlequin and ipsilateral Horner syndromes occurring after unilateral paravertebral anaesthesia for breast cancer surgery. Previously reported incidences of harlequin syndrome in other clinical circumstances and possible mechanisms are discussed.

Case report

A 55-yr-old, 65-kg, ASA II female underwent left mastectomy and immediate latissimus dorsi breast reconstruction for intermediate grade ductal carcinoma. She had a 4-yr history of arterial hypertension, well controlled with an angiotensin II receptor antagonist. After connection of standard monitors and sedation with midazolam 2 mg and fentanyl 50 μg i.v., a left paravertebral block2 was performed, using an 18 G Tuohy needle, at the third thoracic dermatome level with the patient in the right lateral position. Loss of resistance was encountered at 5.5 cm, and a catheter was easily inserted 5 cm into the paravertebral space. Levobupivacaine 0.5% was administered in increments of 5 ml every 5 min to a total of 20 ml. General anaesthesia was then induced with propofol 100 mg and tracheal intubation was facilitated with vecuronium 7 mg. The right radial artery was cannulated for continuous blood pressure monitoring. Diclofenac 100 mg and acetaminophen 1000 mg were given rectally, immediately after induction, for supplementary analgesia. Anaesthesia was maintained for 4 h with sevoflurane in 50% nitrous oxide and oxygen. Two boluses of 5 ml levobupivacaine 0.25% were administered paravertebrally 30 min before the end of surgery. No i.v. opioid analgesics were required. Baseline blood pressure and heart rate were 142/65 mm Hg and 72 beats min⁻¹ respectively. The patient was haemodynamically stable (within 20% of baseline) throughout the whole procedure. A total of 3000 ml i.v. compound sodium lactate was administered. Blood loss was 650 ml.

After operation, the patient received a continuous infusion of levobupivacaine 0.1% at 0.15 ml kg⁻¹ h⁻¹, which was administered via a Perfusor® Compact syringe pump (B. Braun Medical, Sheffield, UK). Pain, assessed on a 100 mm visual analogue scale in the recovery room and every hour after surgery, was zero on movement of the ipsilateral arm. Four hours after surgery, ward staff noticed hemifacial colour asymmetry. In addition to the left ipsilateral classical signs of Horner syndrome (ptosis, miosis, enophthalmos and anhidrosis), there was contralateral right hemifacial flushing and sweating with a well-defined demarcation in the midline. Skin temperature, measured using a 400 Series skin temperature sensor (DeRoyal...
Europe, Kells, Ireland), was 35.5°C on the right side of the forehead compared with 32.9°C on the left. There was no obvious upper limb abnormality of colour and temperature (not measured). Bilateral hand grip and sensation were normal. The patient was fully alert and comfortable and vitals signs were within normal limits. Over the next 6 h, the unilateral facial flushing subsided, the Horner syndrome persisting until the following day.

**Discussion**

Transient unilateral Horner syndrome has been reported after upper thoracic paravertebral block, but, to the best of our knowledge, this is the first report of harlequin appearance occurring with paravertebral anaesthesia.

In 1988, Lance and colleagues first used the term ‘harlequin syndrome’ to describe the sudden onset of unilateral facial flushing and sweating in hot temperatures, extreme emotion or after strenuous exercise in a case series of patients. Harlequin syndrome has been reported subsequently not only as an independent primary complaint in sporting or hard physical work situations, but also associated with other autonomic system disturbances in acquired and congenital Horner syndrome, trauma, stroke and mediastinal tumours. Unilateral facial flushing and sweating may be also encountered in a number of medical conditions, such as idiopathic hyperhidrosis and Frey’s syndrome (gustatory flushing syndrome). The Chinese ethnic group is predisposed to upper body and face redness after alcohol due to genetic variation in alcohol-metabolizing enzymes.

Drummond and colleagues investigated the vasomotor, sudomotor and oculomotor responses in patients with a sympathetic deficit or pharmacologically induced blockade.

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**Fig 1** Autonomic sympathetic fibres distribution to the face. Preganglionic pupillomotor, sudomotor and vasomotor fibres enter the ipsilateral corresponding thoracic ganglia of the sympathetic trunk, through which they ascend into the neck and synapse in the superior cervical ganglion. The postganglionic fibres from the rostral part of the superior cervical ganglion project into the internal carotid nerve to supply the eyes, forehead and cheeks. More caudal postganglionic fibres project into the external carotid nerve to bring the sympathetic supply to cheeks and jaw. (Modified, with permission, from Turco GR, Farber NE. Postoperative autonomic deficit: a case of Harlequin syndrome. *Anesthesiology* 1996; 85: 1197–9.)
of the sympathetic pathway to the face. By exposing these patients to environmental challenges (heat, emotional embarrassment, gustative stimuli), they demonstrated that the conventional sympathetic pathway to the face encompasses sympathetic vasodilator fibres which mediate facial blood vessels dilation in response to heat and emotion, and that a lesion anywhere along the sympathetic pathway to the face impairs thermoregulatory flushing and sweating at the denervated site in response to body heating and emotion.

The upper thoracic sympathetic nervous system can be broken off as white rami communicantes that enter the corresponding ipsilateral thoracic ganglia of the sympathetic trunk, from which they ascend into the neck and synapse in the superior cervical ganglion (Fig. 1). The pupillomotor fibres leave the spinal cord at T1, while the sudomotor and vasomotor fibres exit at T2–T3. The postganglionic fibres from the rostral part of the superior cervical ganglion project into the internal carotid nerve which brings sympathetic innervation to the eyes and forehead. More caudal postganglionic fibres project into the external carotid nerve, providing vasomotor and sudomotor innervation of the jaw. Fibres to the cheeks travel by either route (Fig. 1).

Therefore, the coexistence of Horner and harlequin syndromes in our patient may be attributable to a left-sided pharmacological sympathetic denervation caused by the paravertebral spread of local anaesthetic to the ipsilateral stellate ganglion (formed by the fusion of the first sympathetic thoracic and the inferior cervical ganglion) or preganglionic fibres originating from the first three segments of the thoracic spinal cord (Fig. 1). It has been demonstrated previously that a single dose of bupivacaine 0.5%, 15 ml injected in the thoracic paravertebral space produces unilateral somatic and sympathetic block over five to eight dermatomes.

Harlequin syndrome has been reported only rarely in the perioperative setting. A 9-month-old infant, premature at birth, with a past history of surgical correction of a complex congenital heart disease and tracheostomy, underwent Nissen fundoplication and gastrostomy for recurrent aspiration pneumonia.12 Intraoperatively, the patient developed transient, 1-h duration, well-demarcated plethora of the right neck and face. The authors presented the colour change as a clinical curiosity and no explanation could be attributed to the event. Contralateral hemifacial erythema was also noticed in a 2-year-old child undergoing surgical removal of a cystic hygroma under general anaesthesia and local infiltration of the skin on the operative side.13 The unilateral flushing was exacerbated by crying or coughing. In another case,14 a 3-month-old child developed acquired ipsilateral Horner syndrome secondary to a neuromastoma of the left cervical sympathetic chain. The surgical removal of the tumour was followed by a persistent heat- and crying-exacerbated flushing of the entire body and loss of thermoregulatory vasodilator sympathetic response on the left side of the face, which appeared very pale. The aurotic dysfunction was confirmed by skin conductance response polygraphy. In the above paediatric cases, the harlequin appearance seems to be related to surgical trauma of the cervical sympathetic pathways after difficult dissection of neck masses.

In adults, the only published report of perioperative transient harlequin syndrome was associated with the uneventful first-pass cannulation of the right internal jugular vein in a patient undergoing laparotomy for an intra-abdominal collection.15 No local anaesthetic infiltration was used. The patient had previously had right internal jugular vein cannulation and subsequent removal of a Hickman catheter. Neuropraxia of the cervical sympathetic supply to the face during repeated right neck cannulation was proposed as an explanation.

In conclusion, we observed contralateral facial flushing with ipsilateral Horner syndrome after paravertebral anaesthesia. This was due to a normal or excessive vasodilatory, thermoregulatory response to heat or emotion by an intact sympathetic pathway on the erythematous (right) side together with relative pallor of the left side because of high thoracic sympathetic blockage. The occurrence of harlequin syndrome after paravertebral anaesthesia appears to be transient and requires only explanation and reassurance.

References