Postoperative Autonomic Deficit: A Case of Harlequin Syndrome

Glenn R. Turco, D.O.,* Neil E. Farber, M.D., Ph.D.*

HARLEQUIN syndrome in adults was first described by Lance et al.¹ as the sudden onset of unilateral facial flushing and sweating. The term “harlequin color change” has been used for years to characterize newborns with unilateral flushing of the dependent half of the body.²–⁵ Although the etiology of these unilateral color changes is unknown, it appears to require a deficit or dysfunction of the autonomic nervous system, and to result in vasomotor instability.⁶–⁹ There have been no previous reports of perioperative harlequin syndrome or unilateral color disturbances. We present a case of postoperative, well-demarcated, left hemifacial flushing and sweating after resection of a right neck mass in a child.

Case Report

A 2-yr-old, American Society of Anesthesiologists physical status I child, who weighed 12 kg, presented for elective resection of a neck mass. The mass was 2-3 cm, mobile, nontender, located in the anterior cervical portion of the right neck, and was noted to be increasing in size throughout the last 5 or 4 months. A computed tomography scan revealed a 2 × 3 × 1.5 cm solitary cystic mass medial to the lateral head of the sternocleidomastoid muscle, associated with the neurovascular structures. There were no masses in the anterior mediastinum or airway impingement. The neck mass was palpable in the anterior cervical and submandibular right neck and was thought not likely to interfere with laryngoscopy. The patient was clinically asymptomatic, had a Mallampati classification of I, and a medical history that was otherwise unremarkable.

General anesthesia was induced uneventfully with halothane in nitrous oxide and oxygen. The trachea was intubated after administration of 1.0 mg·kg⁻¹ rocuronium, and anesthesia was maintained with 0.5–1% halothane in a 2:1 mixture of nitrous oxide and oxygen. We administered 0.1 mg·kg⁻¹ morphine sulfate for perioperative analgesia. The patient was positioned supine. To facilitate surgical exposure, the neck was extended by placement of a paddled roll under the scapula, which elevated the shoulders 2 cm. The head rested freely on a foam doughnut and was turned 70° to 90° to the left. Before incision, the skin was infiltrated with 4 ml of 1% lidocaine + 1:200,000 epinephrine. As the operation proceeded, the mass was noted to be adherent to the internal jugular vein, carotid artery, and the vagus nerve anterolaterally and was excised by blunt and sharp dissection. The mass was thought to be a benign vascular lesion, and later histologic studies suggested a cystic hygroma. The intraoperative course was uneventful, with stable respiratory and hemodynamic status throughout dissection, except for one 10 s decrease in heart rate from 110 to 80 beats per minute, without rhythm change and requiring no intervention because of its transient nature.

At the conclusion of surgery, neuromuscular blockade was antagonized with 0.07 mg·kg⁻¹ neostigmine and 0.015 mg·kg⁻¹ glycopyrrolate. After removal of the surgical drapes, a sharp midline facial demarcation (fig. 1) was observed. The face contralateral to the operative site was flushed, warm, and wet with perspiration, and these signs were noted to increase in intensity during coughing or coughing. The ipsilateral operative side, in contrast, was pale, cold, and dry to touch, and its color did not change with crying or coughing. The pupils were miosis and equal and reactive to light. The surgical site had been decompressed previously via a percutaneous drain, and, on inspection, no excessive bleeding or drainage was seen. The child appeared to be ventilating well before and after tracheal extubation. The patient was taken to the postanesthesia recovery unit and, after an uneventful course, with minimal improvement in the facial coloration, was discharged to the short stay unit. At this time, it was noted that facial nerve function was grossly intact bilaterally, that no palsy was present, and that the nasal mucosa was not congested on either side. During an additional 3-h period, the facial flushing and sweating had resolved completely, and the patient was discharged to home.

During follow-up on postoperative days one and ten, there was no sign of the red facial discoloration observed perioperatively, nor was there evidence of autonomic, motor, or sensory neuropathic deficit.

Discussion

There are several autonomic syndromes associated with unilateral facial flushing. Along with the distribution of flushing, the presence or absence of associated autonomic signs and symptoms may be used to localize the primary
CASE REPORTS

Fig. 1. A patient with hemifacial discoloration 30 min postoperatively in the recovery room.

Lesion along a three-neuron pathway. Central sympathetic lesions may occur with brainstem infarction or with migraine, producing a central Homer's syndrome or a Horner's syndrome accompanied by hemi-body loss of thermoregulatory sweating.\(^{16-18}\) Alternatively, immaturity of the hypothalamic centers may be responsible for the unilateral flushing often seen in premature neonates that is termed Harlequin color change.\(^ {1,2}\) Second order preganglionic oculomotor fibers leave the spinal cord in the first thoracic root,\(^ {1,2}\) whereas sudomotor and vasomotor fibers leave below T1, at T2 and T3. Noda\(^ {7}\) described a patient with Harlequin syndrome due to a superior mediastinal mass, interfering with second order neurons at T3. The preganglionic fibers then ascend via the sympathetic chain to synapse in the superior cervical ganglion (fig. 2). Postganglionic neurons in the rostral end of the superior cervical ganglion project into the internal carotid nerve to innervate the eyes and forehead, whereas fibers from the caudal end of the superior cervical ganglion project into the external carotid nerve to innervate the cheeks and other parts of the face.\(^ {1,2}\) The upper extremity receives postganglionic fibers from the stellate ganglion, so a lesion in or proximal to the stellate ganglion would disrupt sudomotor and vasomotor responses to the arm. In the case presented, upper extremity sudomotor or vasomotor changes did not occur, delineating the site of sympathetic injury as distal to the stellate ganglion. Nor did we observe oculomotor changes (ptosis or meiosis), suggesting that the injury occurred to preganglionic fibers traveling to the superior cervical ganglion arising from T2 to T4, to the caudal superior cervical ganglion, or to the nerves of the external carotid plexus distal to the superior cervical ganglion. In addition, the likeness of the superior cervical ganglion to an enlarged lymph node can result in its inadvertent removal or injury during neck dissection, producing a Horner's syndrome or autoneuropathy.\(^ {19}\) The extensive dissection required, both medially and posteriorly to the carotid artery to resect the cystic hygroma, made it more likely that autonomic injury would occur, and may explain why this phenomenon had not been described after a relatively superficial neck surgery like carotid endarterectomy.

The determination of the pathologic side of the face is somewhat ambiguous. As the dominant effect of the sympathetic nervous system to the face is one of vasodilatation, reactive vasodilatation may develop immediately after sympathectomy. This may imply that the sympathetic abnormality in the current patient would be on the nonsurgical side and may have resulted from extreme neck rotation leading to ischemia of the left sympathetic nerves. However, facial flushing is a reflection of the vasodilatation of cutaneous blood vessels, which may be caused by attenuation of sympathetic vasconstrictor tone, by an active sympathetic vasodilator mechanism, or by endogenous or exogenous vasodilators.\(^ {5}\) The relative importance of the sympathetic influence on facial blood flow was studied by Drummond\(^ {11}\) in patients undergoing stellate ganglion blockade (preganglionic sympathetic fibers to the superior cervical ganglion will also be blocked). The release of sympathetic vasconstrictor tone increased ipsilateral facial blood flow by only 40%. In contrast, heating until sweating was visible on the normal side of the forehead increased blood flow 150 to 200%, with no change in blood flow to the sympathetically blocked side. The reciprocal effect of sympathetic hyperactivity in the forehead region is poorly understood, but it has been demonstrated that forearm vasodilation can occur without thermal stimulation.\(^ {20}\)


Anesthesiology, Vol 85, No 5, Nov 1996
Fig. 2. Autonomic sympathetic innervation of the face. The preganglionic supply of the eye exit the spinal cord in the first thoracic root, whereas preganglionic sudomotor and vasomotor fibers leave below T1. The rostral superior cervical ganglion projects postganglionic fibers to the eyes, forehead, and cheeks via the internal carotid nerve, whereas the caudal part of the ganglion projects postganglionic fibers to the cheeks and other parts of the face via the external carotid nerve. Numbers 1, 2, and 3 represent sympathetic lesions that may produce hemifacial loss of flushing with ocular sparing. 1 = preganglionic fibers from T2 and T3; 2 = caudal superior cervical ganglion; 3 = postganglionic fibers of the external carotid nerve.

References