Delayed Onset of Harlequin Syndrome after Vascular Ring Repair

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Harlequin syndrome was first described in the literature by Lance et al in 1988 in five adults with sudden onset unilateral facial flushing and sweating. Triggers include thermal, gustatory, emotional, or exertional stimuli. This rare syndrome has been described independently in hard physical work situations, autonomic dysfunction associated with congenital Horner’s syndrome, stroke, trauma, internal vein catheterization, neck mass resection, and regional anesthesia.1-9

We present a case of a 4 yr old, 22.5 kg, ASA II, male child who presented for division of a vascular ring. He had experienced recurrent episodes of wheezing throughout his life. Further workup revealed a diagnosis of a vascular ring with right aortic arch and aberrant left subclavian artery with resultant tracheal compression.

In the OR, general anesthesia was induced with sevoflurane in nitrous oxide and oxygen. Tracheal intubation was facilitated with rocuronium and anesthesia was maintained with sevoflurane in an air and oxygen mixture. Following right lateral decubitus positioning, the surgeon performed a left posterolateral thoracotomy incision to enter the chest through the 4th intercostal space. The mediastinal pleura was opened and the vascular ring divided by ligation and transection of the ligamentum arteriosum.

An intercostal nerve block at the T3-T5 levels was performed by the surgeon using 10 ml of 0.25% bupivacaine with 1:200,000 epinephrine for postoperative analgesia. Upon successful completion of the surgery, the patient was extubated and taken to the PACU in stable condition.

Approximately 3 hours after the nerve block, the right side of the patient’s face was noted to be markedly flushed and perspired with a well-defined midline demarcation. This stood in stark contrast to the pale, anhidrotic left side of the face ipsilateral to the surgical site. No other neurologic abnormalities were noted. Specifically, no ptosis or miosis was present. The flushing and perspiration subsided within 3 hours from onset. The patient recovered well from surgery and was discharged home 2 days later with no recurrence of similar symptoms.

The pathophysiology of harlequin syndrome is believed to result from unilateral autonomic interruption of the upper thoracic segments of the sympathetic chain, responsible for vasomotor fibers supplying the face. Anesthetic blockade of these fibers results in reduced ipsilateral skin blood flow. Horner’s syndrome may be found in conjunction with harlequin syndrome. The oculcar findings of Horner’s are likely related to blockade at the level of T1 while the sudomotor and vasomotor findings of harlequin syndrome are thought to be associated with the levels T2 and T3. Onset of symptoms range from 30 minutes to 4 hours according to case reports.4,7,9 Resolution of ocular symptoms may persist for greater than 12 hours while sudomotor and vasomotor symptoms resolve within 2 to 6 hours after onset.5,6,7,9 There have been no reported long-term neurological sequelae.

References:


* Used with permission from Turco GR, Farber NE. Postoperative autonomic deficit: a case of harlequin syndrome. Anesthesiology 1996;85:1197-1199.


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