Interscalene brachial plexus blockade is regularly used for postoperative pain management following shoulder surgery. A known but generally benign side effect of this technique is Horner syndrome. Another syndrome known as harlequin syndrome exists but does not appear to be as common. This syndrome consists of contralateral facial flushing and sweating secondary to ipsilateral sympathetic chain inhibition. Despite the alarming presentation in the perioperative setting, this syndrome appears to be benign and self-limiting when precipitated by regional anesthetic technique. This article describes an occurrence of harlequin syndrome without observed ptosis or miosis following a postoperative interscalene nerve block.

Keywords: Brachial plexus, harlequin syndrome, interscalene, regional anesthesia, sympathetic chain.

Isolated Harlequin Syndrome Following Brachial Plexus Nerve Block via Interscalene Approach: A Case Report

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The interscalene approach to brachial plexus blockade is commonly used to provide postoperative analgesia for shoulder or arm surgery. Although Horner syndrome is a well-documented complication of this technique, harlequin syndrome is not. Harlequin syndrome consists of a unilateral facial flushing and sweating. There is a distinct vertical line of demarcation usually extending the length of the face producing a rather profound appearance. The pathogenesis of harlequin syndrome is highly variable and can even be idiopathic in nature.

Documented reports of harlequin syndrome have occurred following thoracic paravertebral epidural placement, multilevel intercostal block, and internal jugular catheterization. Our case was consistent with the description by Lance et al., who conceived the term harlequin syndrome, as a dysfunction of vasomotor innervation with a lack of ipsilateral flushing and sweating.

Case Summary

A 60-year-old female, ASA class 3, presented for an arthroscopic left rotator cuff repair. She weighed 119 kg and was 162.5 cm tall. Her allergies were amoxicillin, azithromycin, ciprofloxacin, clarithromycin, and sulfa antibiotics. The patient’s medical history was remarkable for type 2 diabetes, hypertension, hyperlipidemia, reflux with esophagitis, severe postoperative nausea and vomiting requiring hospital admission, and a rotator cuff tear. She also had Paget disease of the vulva requiring biopsies, groin lymph node removal, and ultimately partial removal of the vulva.

The patient gave consent for both general and regional anesthesia but declined to have a peripheral nerve block done in the preoperative period, with the hope of avoiding the procedure altogether. The patient was preoperatively given oral acetaminophen, pregabalin, and tramadol. A scopolamine patch was placed postauricularly. Two milligrams of midazolam was administered intravenously for reduction of anxiety before the patient was transported to the operative suite. General anesthesia was induced with 100 µg of fentanyl, 100 mg of lidocaine, and 200 mg of propofol, and endotracheal intubation was facilitated with succinylcholine, 140 mg, which proceeded uneventfully. Rocuronium, 30 mg, was administered for muscle relaxation, followed by 20 mg of famotidine and 10 mg of dexamethasone. The arthroscopic rotator cuff repair was successfully completed, with the bed turned 45 degrees clockwise in the supine position, as opposed to the lateral decubitus or sitting position. Five-lead electrocardiogram, ventilation, peripheral oxygen saturation, noninvasive blood pressure, and temperature were monitored throughout the procedure. The patient remained hemodynamically stable throughout the procedure.

After completion of the procedure, the patient was given 4 mg of intravenous ondansetron, and 3 mg of neostigmine with 0.6 mg of glycopyrrolate for reversal of neuromuscular blockade. She was then successfully extubated and transported to the postanesthesia care unit, where she received an additional 50 µg of fentanyl.

The anesthesia team was recalled to the bedside in the recovery area approximately 30 minutes later because the patient requested peripheral nerve blockade. The patient was again sedated with 2 mg of midazolam. Following a procedural time-out and establishment of sterile technique, a left-sided interscalene block was initiated using a nerve stimulator. Although a biceps response was elicited...
successfully, this occurred at a location much more posterior than initially anticipated. A total of 23 mL of local anesthetic was injected, and aspiration was performed every 5 mL. The local anesthetic used was a mix of 15 mL of 0.5% ropivacaine and 10 mL of 1% tetracaine.

An hour following the nerve block, the anesthesia team was again called to the recovery area to evaluate the patient. On examination, the patient was found to have right-sided facial flushing and sweating, contralateral to the left-sided block. The patient did not have ptosis or miosis. She denied facial pain or numbness. The patient recovered from anesthesia adequately to meet discharge criteria and was discharged home. During telephone follow-up the next day, the patient reported no complaints and reported that the harlequin sign had disappeared.

Discussion
There have been publications regarding harlequin syndrome following other regional techniques; however, a thorough review of the literature failed to return a case report of harlequin syndrome following interscalene brachial plexus blockade. During the process of researching and writing this article, conversation with other practitioners revealed that in the presence of Horner syndrome, harlequin syndrome may go overlooked. Therefore, it may be possible that the occurrence of harlequin syndrome is more frequent in regional anesthesia than published reports indicate.

Even though the pathogenesis of harlequin syndrome is variable, it can be reasonably deduced in the case of regional anesthesia that it is related to the inhibition of sympathetic chain outflow by local anesthetic infiltration.9 A case report by Sribnick and Boulis10 demonstrated complete resolution of an iatrogenically caused harlequin syndrome in a patient who presented without the attributes of Horner syndrome. The authors accomplished this by a surgical sympathectomy of the functional T2 ganglion. This reinforces that the second thoracic ganglion is implicated in isolated harlequin syndrome, whereas stellate ganglion lesions or blockade induce Horner syndrome.9 Nonetheless, it is a pathophysiologic curiosity when harlequin syndrome presents without Horner syndrome attributes in the setting of interscalene brachial plexus blockade. The question that arises is whether our patient had an anatomical anomaly or whether the volume of local anesthetic infiltration was able to somehow reach both the brachial plexus and the T2, but not the stellate, ganglion. It is possible that because of the posterior location of the needle and the patient’s body habitus, a combined brachial plexus and partial paravertebral block was performed inadvertently.

Fortunately, harlequin syndrome is usually a benign and self-resolving complication of regional anesthetic technique. A case by Majumder et al4 confirms the resolution of a harlequin syndrome, isolated from Horner syndrome, approximately 9 hours after a T3/T4 paravertebral block was performed. Boling et al11 describe a case of harlequin syndrome in 2014 resulting from an epidural infusion catheter at the T7 level, which was threaded 5.5 cm into the epidural space. The authors withdrew the catheter to 3 cm in the epidural space and paused the infusion for 3 hours. The harlequin sign resolved overnight. Two cases of harlequin syndrome presented by Thomas and Polaner12 were secondary to thoracic epidural analgesia in pediatric surgical patients. On decreasing or stopping the epidural infusion and repositioning the catheter, both cases resolved within hours. In a recent nonanesthetic case of interest, Fringeli et al13 describe coexisting harlequin and Horner syndromes due to the compression of the cervical sympathetic chain by a surgical drain following anterior cervical disectomy. On removal of the offending drain, the harlequin sign resolved within a few hours.

It is possible to resolve harlequin syndrome by performing a paravertebral T2 sympathetic ganglion block. An article by Reddy et al14 investigated the treatment of harlequin syndrome by blocking the unaffected sympathetic chain with local anesthetic. This treatment proved initially successful but was limited by patient anxiety to repeated procedures. Although local anesthetic is an effective measure to attenuate the facial flushing of harlequin syndrome, when it is induced by regional anesthesia, it is much simpler and safer to educate and reassure the patient, and wait for metabolic resolution of the syndrome. In regional anesthesia, it is evident that once sympathetic chain inhibition ceases, harlequin syndrome resolves.

REFERENCES


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**DISCLOSURES**

The authors have declared no financial relationships with any commercial entity related to the content of this article. The authors did not discuss off-label use within the article.