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CASE REPORT

Late Postoperative Harlequin Syndrome Coexisting With Horner Syndrome After Thoracic Epidural Anesthesia

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A patient who underwent left mastectomy under thoracic epidural anesthesia developed transient ipsilateral well-demarcated hemifacial erythema, sweating, and contralateral Horner syndrome postoperatively. All the clinical features subsided and had completely resolved within 24 hours after the cessation of patient-controlled epidural analgesia with bupivacaine 0.05% and fentanyl 0.5%. We believe that the reason for the excessive high-level thoracic sympathetic blockade that led to this concomitant syndrome was cephalad spread of local anesthetic.

1. Introduction

The predominant features of Harlequin syndrome are unilateral facial flushing and sweating. Harlequin syndrome has been reported in different clinical conditions including brain stem infarction, superior mediastinal neurinoma, and internal jugular vein catheterization.[1,2] Idiopathic and iatrogenic cases have been reported. The clinical features of Horner syndrome are ptosis, miosis, enophthalmos and anhidrosis. Horner syndrome is generally reported after trauma, stroke and mediastinal tumors, due to autonomic system disturbances.[3–5] A few cases of coexisting Harlequin and Horner syndromes due to anesthesia have been reported in the literature. Burlacu and Buggy reported a transient coexisting unilateral Horner and Harlequin syndromes after upper thoracic paravertebral block in combination with general anesthesia.[6]

We present a patient who had coexisting Harlequin and Horner syndromes after thoracic epidural anesthesia (TEA) for left modified radical mastectomy, and discuss the clinical features and correlation with TEA.

2. Case Report

A 47-year-old woman (weight, 62 kg; height, 152 cm) with carcinoma of the left breast (T3N0M0) was scheduled for modified radical mastectomy. She had euthyroid multinodular goiter and had been on progesterone therapy for atrophic endometritis with intermittent vaginal bleeding for the last 3 weeks. Preoperative evaluation revealed no remarkable findings.

TEA was planned for perioperative anesthesia and analgesia. After obtaining patient consent for surgical

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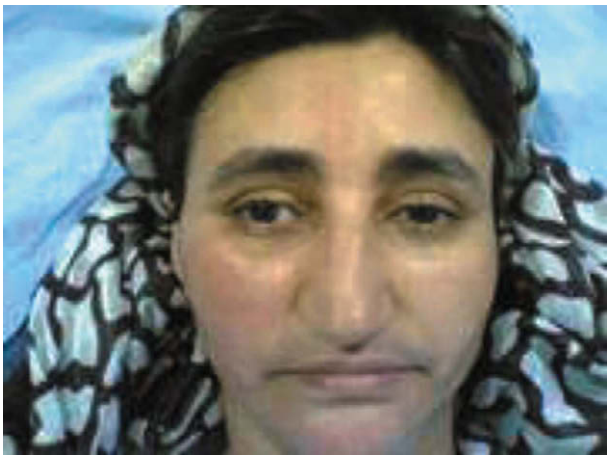

Figure 1 Coexisting transient Harlequin and Horner syndromes after thoracic epidural anesthesia for modified radical mastectomy in the postoperative period. Figure is reprinted with permission from Poster no. 291: Aydin T, Sahin L, Algin C, Yaylak F, Hacioglu A. Transient coexisting Horner and Harlequin syndromes after thoracic epidural anesthesia for modified radical mastectomy. *Supplement to Regional Anesthesia and Pain Medicine, XXVI Annual ESRA Congress, Valencia, Spain.* 2007; 32(5 Suppl 1):46. Copyright Elsevier (2007).

technique and TEA, an epidural catheter (20 G Portex disposable epidural catheter) was placed into the T3–4 interspace through an 18G Touhy needle, using the median approach and loss-of-resistance technique with the patient in a sitting position. The catheter was directed 3–4 cm cephalad position. A total of 10 mL of bupivacaine 0.5% was administered in two sequential doses at 5-minute intervals. No additional analgesic and sedative medication was used until the end of surgery. The operation was completed uneventfully and the patient was transferred to the recovery room with a patient-controlled analgesia (PCA) pump. The PCA solution was prepared with 20 mL bupivacaine 0.5% and 5 mL fentanyl 0.5% diluted in 225 mL physiologic saline. The infusion rate was set at 0.13 mL/kg/hr, and the bolus dose was set at 0.1 mL/kg with a 4-hour lock-out interval.

Three hours after surgery, right hemifacial color asymmetric flushing and sweating with left-sided classical signs of Horner syndrome (ptosis, miosis, enophthalmos and anhidrosis) with a well-defined demarcation in the midline was observed (Figure 1). No upper limb abnormality in terms of color and temperature was present. Bilateral handgrip and sensation were normal. The patient was fully alert and comfortable. Vital signs were within normal ranges. PCA infusion was stopped immediately and the epidural catheter removed. The total dose delivered to the patient was 8.06 mL at that time. Unilateral facial flushing subsided 6 hours after the termination of bupivacaine infusion. The Horner syndrome on the left persisted to the following day. Analgesia was continued with patient-controlled intravenous morphine analgesia. The regimen was adjusted to infuse 1 mg/hr of morphine with a lock-out interval of 10 minutes.

3. Discussion

Harlequin syndrome is a sympathetic deficit that is generally confined to the face on the non-flushing side. Rarely, autonomic deficit may involve the parasympathetic neurons in the ciliary ganglia.[6,7] Harlequin syndrome was first described by Lance et al as a sudden onset of unilateral facial flushing and sweating.[3] Iatrogenic Harlequin syndrome has been reported.[6]

Upper thoracic sympathetic fibers carry afferent stimuli to the ipsilateral thoracic ganglia, which communicates with the superior cervical ganglia of the sympathetic nervous system. Efferent stimuli from the superior cervical ganglia are carried with the fibers of the internal and external carotid nerve. The internal carotid nerve carries sympathetic stimuli to the eyes and forehead, whereas the external carotid nerve carries sympathetic stimuli to the cheeks and the jaw.[8]

Burlacu and Buggy were the first to report coexisting Harlequin and Horner syndromes after modified radical mastectomy and immediate breast reconstruction performed under combined paravertebral block (with a total of 15 mL levobupivacaine 0.25%) and general anesthesia.[6] Saito also demonstrated unilateral somatic and sympathetic block over five to eight dermatomes produced with thoracic paravertebral injection of a single dose of 15 mL bupivacaine 0.5%.[7] In contrast to Burlacu and Buggy, we chose TEA because of its beneficial effects on the respiratory and cardiac systems and its efficiency in providing preoperative analgesia.

The appearance of Horner syndrome is not necessarily associated with signs of sensory or motor blockade at the cervical or upper thoracic dermatomes. We believe that cephalad spread of the local anesthetic solution blocked the transmission of efferent sympathetic activity to the pupillomotor, sudomotor, and vasomotor fibers at T1–3 levels, respectively. In addition, this theory explains the occurrence of coexisting Harlequin and Horner syndromes after TEA in our case. The time of occurrence of the symptoms and the position of the patient may help to answer the question of why these syndromes occurred 3 hours after surgery. Epidural anesthesia was performed by an experienced anesthetist with the patient in a sitting position. Surgery was performed with the patient in supine position, and patient was kept in the semi-Fowler position both in the recovery room and in the ward. We speculate that a change in the axis of the cervical column due to changing the position of the patient from supine to the semi-Fowler position might have caused migration of the epidural catheter, leading to a high level of thoracic sympathic blockade. In order to prevent this situation, we should have checked the level of epidural anesthesia after the change in patient position, but we did not. However, the course was uneventful and the patient recovered completely.

In conclusion, transient contralateral Harlequin and ipsilateral Horner syndrome after TEA may be due to normal or excessive contralateral side plus ipsilateral high-level thoracic sympathetic blockade.

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