Postoperative Harlequin Syndrome:
Case Report of a Rare but Clinically Striking Condition
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SUMMARY:
- Two year old boy undergoing thorascopic resection of a left paraspinal mediastinal mass
- Postoperatively developed harlequin syndrome
- Discussion highlights this unusual condition and the anesthetic concerns related to the differential diagnosis presenting similarly postoperatively

CASE REPORT:
- Newly diagnosed left paraspinal mediastinal mass, most consistent with neurablastoma, in an otherwise healthy 2 year old boy
- General anesthesia for thorascopic resection
- Standard ASA monitors and 22G left radial arterial line inserted intra-op
- Patient positioned in right lateral decubitus position, with an axillary roll and pressure points well padded with extremities in neutral position
- Uneventful case and patient extubated without complication
- On arrival to PACU, patient noted to have an erythematous rash on the right side of his face
- Initial impression was the rash was a result of exposure of the patient's fair, sensitive skin to the forced air warming device
- After about an hour, the rash had become more delineated and progressed to the upper right chest and arm, with a distinct line directly midline
- After conversation with the surgical team, a diagnosis of harlequin syndrome was made

DISCUSSION:
Harlequin syndrome is a rare condition, affecting less than 200,000 people in the United States. It is characterized by unilateral flushing of the face, neck and upper thorax with a sharp midline demarcation. The ‘Harlequin Sign’ (unilateral flushing with or without sweating) is occasionally noted after strenuous activity, exposure to heat or secondary to emotional triggers. The etiology of harlequin syndrome is thought to be a lesion of the preganglionic or postganglionic cervical sympathetic fibers and parasympathetic neurons of the ciliary ganglion. The sympathetic deficit on the denervated side results in flushing of the contralateral side. It is unclear whether the response of the undamaged side is normal or excessive, but it is believed to be the result of a compensatory mechanism for damage to the affected side. In our patient, the etiology was attributed to a lesion of the sympathetic chain ganglia at T6, caused during resection of the patient’s left T2-T6 paraspinal tumor.

DIFFERENTIAL DIAGNOSIS:
- Neurovascular diseases (such as acute stroke or intracranial hemorrhage) involving the autonomic nervous system
- Malignancy of the brain or lung
- Venous or arterial thrombus
- Trigeminal neuralgia, Frey’s syndrome, or Horner’s syndrome
- Horner’s syndrome also known to present in conjunction with harlequin syndrome
- Also transient etiologies of harlequin syndrome can be induced by complications of anesthesia including:
  - Neuropraxia of the sympathetic innervation of the face after IJ catheterization
  - Administration of extrapleural bupivacaine infusion
  - High thoracic paravertebral anesthesia
  - Asymmetrical epidural anesthesia

REFERENCES: