Right VATS Sympathectomy For Treatment Of Harlequin Syndrome; A Case Report

Dr F Kirk, Dr K Crathern & Dr A Stroebel

Introduction

1st Described in 1988, named from English folklore (1, 2)
Harlequin Syndrome is characterized by Exercise/Stress/Temperature induced;
- Unilateral facial flushing and Hyperhidrosis
- With contralateral anhidrosis (Figure 1)

Causes:
- Benign physiology of neonates
- Immunological/ Multiple sclerosis
- Cerebrovascular Disease/stroke
- Neoplastic (benign or malignant) i.e. apical lung Ca, Spinal cord tumour, Goitre (3,4)
- Paravertebral (transient) & rarely thoracic epidurals

Pathophysiology

Harlequin syndrome is believed to be the result of dysregulated/excessive sympathetic nervous stimulation of the blood vessels and dermal sweat glands of the face. The facial SNS outflow moves from the thalamus through the brainstem, spinal cord to the sympathetic chain at the level of T_{2-3} before ascending to the face via the superior cervical ganglion along the carotid to the face. (1) Any lesion at any point in this outflow tract can produce Harlequin syndrome as described in figure 2



Figure 1: Pre-operative photograph of Harlequin Syndrome

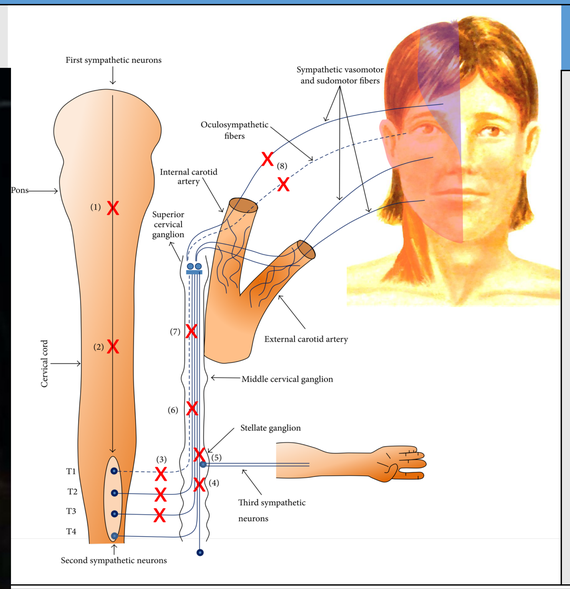


Figure 2: Sympathetic Outflow

Figure 2: Diagram demonstrating the sympathetic innervation to the face with possible lesions sites.(1) pons. (2)the spinal cord. (3) thoracic roots T1-3. (4)= sympathetic chain between T1 and T2. (5)= stellate ganglion. (6)= sympathetic chain between stellate and middle cervical ganglion. (7) = sympathetic chain between middle and superior cervical ganglion. (8)= sympathetic fibres traveling with the internal carotid. Source: Algahtani et. Al 2017 (1)

Case Presentation

- 33-year-old Female received an L4-L5 epidural anaesthetic for child-birth
- Delivery was uncomplicated
- Post-partum was treated empirically with Abx for fever
- Later returned with Retained products + spinal anaesthesia
- Developed Persistent Harlequin syndrome (following discharge)
- MRI Angiography of the head and spine was unremarkable
- Nil intervention by Neurology
- Referred for sympathectomy at 39 years

Management: Right VATS T_2-T_3 Sympathectomy

- 2 port technique & 5mm Camera
- Single lung ventilation &CO_2 Insufflation
- Sympathetic Chain at T_2 & T_3 divided with Harmonic Scalpel
- Lung inflated & CO2 drained with underwater seal technique.
- Ports closed without placing and ICC

Immediate and sustained (9 months) resolution of Harlequin Syndrome

Discussion

Harlequin Syndrome is noted as a temporary complication, after paravertebral anaesthesia. Here it was believed to be the result of one of the anaesthetics (spinal or epidural). However, retrospectively this is contentious as an epidural and spinal anesthetic should produce bilateral, transient signs, rather than persistent unilateral disease. Due to the heterogeneity and rarity of the syndrome little is known about its treatment. Oxybutynin with propranolol and botulin injections have been described, with varying efficacy (5). Described surgical interventions are a costotransversectomy and concurrent sympathectomy, effectively treating the Harlequin syndrome, however produced T1 radicular pain (6). Division of the Sympathetic Chain below the Stellate Ganglion should avoid Horner's Syndrome (preserving T1). Here we describe curative right VATS sympathectomy for Harlequin Syndrome, without radicular pain or complication.

References

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