Harlequin syndrome and Horner syndrome after neck schwannoma excision in a pediatric patient

A case report

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Abstract

Rationale: Harlequin syndrome is an extremely rare benign condition characterized by unilateral facial flushing and sweating. The etiology is not well defined and the pathophysiology is related to dysfunction of the upper sympathetic nerve. Herein, we report a case of harlequin syndrome associated with Horner syndrome in an 11-year-old boy who underwent excision of right neck schwannoma.

Diagnosis: The preoperative diagnosis was neurogenic tumor of vagus nerve or sympathetic nerve.

Interventions: We performed right neck mass removal under general anesthesia.

Outcomes: We report a case of harlequin syndrome associated with Horner syndrome in an 11-year boy who underwent excision of right neck schwannoma.

Lessons: Clinicians should consider the point that harlequin syndrome could occur as a first sign of more serious conditions.

Abbreviation: CT = computed tomography.

Keywords: harlequin syndrome, Horner syndrome, schwannoma

1. Introduction

Harlequin syndrome is an extremely rare neurological condition characterized by unilateral facial flushing and sweating. The etiology is not well defined and the pathophysiology is related to dysfunction of the upper sympathetic nerve. Herein, we report a case of harlequin syndrome associated with Horner syndrome in an 11-year-old boy who underwent excision of right neck schwannoma.

2. Case report

An 11-year-old boy presented with complaint of a right neck mass of 1-month duration. The patient’s medical history was unremarkable. The patient denied fever, chills, pain, or tenderness. Upon physical examination, a firm, 3.0 cm sized mass was palpated in the right upper lateral neck. Neck computed tomography (CT) scans demonstrated a 3 x 4 cm sized oval, well-defined, heterogeneously enhancing mass with internal necrotic foci in the right carotid space. The right neck mass was performed: however, the adequate cell harvest was a failure because of the hardness of the mass. Based on these observations, the preoperative diagnosis was neurogenic tumor of vagus nerve or cervical sympathetic nerves.

3. Discussion

Harlequin syndrome is usually idiopathic; however, in some cases it has been reported to be associated with underlying lesions, such as brain stem infraction, tumor, surgery, or surgical procedures. In the case of our patient, harlequin syndrome and Horner syndrome were contemplated to have occurred by cervical sympathetic nerves injury and concomitant damage of the superior cervical ganglion during surgery.

Harlequin syndrome can also be the first sign of more serious conditions, therefore, diagnostic examinations are required.
Clinicians should refer to patient’s medical history, such as previous tumor, surgery, or trauma. Clinical and neurologic examinations should be performed to reveal the site of the sympathetic lesion. Radiologic examinations, such as CT, magnetic resonance imaging, ultrasound, and electrophysiological procedures should be performed to exclude the presence of an underlying disease. In the present case, we only performed clinical and neurologic examinations without imaging techniques, as the condition was a definite cause of harlequin and Horner syndrome.

Harlequin syndrome is usually benign in nature, and does not need any specific treatment. Reassurance and avoidance of any aggravating factors should be provided to patients. In the case of our patient, harlequin syndrome spontaneously resolved without any treatment.

In conclusion, harlequin syndrome is an extremely rare benign condition, and can occur by excision of neck tumor, even in a pediatric patient. Clinicians should consider the point that harlequin syndrome could be the first sign of more serious conditions.

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