Harlequin syndrome in children: secondary until proven otherwise

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DESCRIPTION
Harlequin syndrome is a rare autonomic disorder characterised by well-delimited unilateral hyperhidrosis and flushing. Vascular smooth muscle and eccrine sweat glands are controlled by the sympathetic nervous system, whose fibres originate from the hypothalamus, descend in the brainstem and exit the spinal cord at T1 to T3 level. Depending on the affected part of this pathway, Harlequin syndrome can have different aetiologies: hypothalamic immaturity, lesion of the sympathetic fibres by compression or surgical or anaesthetic procedures; or idiopathic, triggered by exercise, heat or emotions. Although most adult cases are idiopathic, a great proportion of the reported cases in children have secondary causes.

A boy in early childhood was referred to the paediatric neurology outpatient clinic for a single episode of unilateral facial and upper body sweating during a febrile illness. His mother brought photographs that evidenced the sharply demarcated right-side cranial sweating (figure 1). The event never repeated itself, even at other febrile episodes. He had an uneventful medical and surgical history and didn’t take any medications. There were no other associated symptoms or signs, including headaches or sensory or motor deficits, his vital signs were in the normal range and his physical examination, including a comprehensive neurological assessment, was unremarkable. Laboratory investigation, comprising thyroid function, and cerebral, spine and thoracic magnetic resonance angiography excluded secondary causes. Considering the diagnosis of idiopathic Harlequin syndrome, the benign nature of the condition was explained to the child and his parents, and they continued to follow-up with his general paediatrician.

Harlequin syndrome presents with a characteristic sudden onset hemifacial pallor and anhidrosis, with a compensatory flushed andhidrotic contralateral side. The changes can also comprise the arms and chest, depending on the extension of the nerve fibres affected, or involve the entire half of the body, especially in neonates. Vasomotor instability due to hypothalamic immaturity is very common in newborns and it is responsible for the high incidence of Harlequin syndrome in this population. Autonomic nerve fibre compression can be caused by goitre, thoracic tumours or carotid artery dissection, and neurological diseases such as Guillain-Barré syndrome or multiple sclerosis can also present with these findings. A detailed history and careful physical examination, as well as various analytical and imaging studies, are essential to rule out underlying conditions. Idiopathic Harlequin syndrome is a benign self-limiting condition requiring reassurance to the patient and family about its autoresolutive nature.

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Learning points
► Harlequin syndrome is a rare neurological disorder with multiple possible aetiologies.
► Secondary serious causes are more frequent in paediatric patients.
► Awareness of this syndrome is crucial for appropriate patient care.

Figure 1 Demarcated right-side cranial hyperhidrosis.
Patient consent for publication  Consent obtained from parent(s)/guardian(s).

Provenance and peer review  Not commissioned; externally peer reviewed. Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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