The Journal of Emergency Medicine, Vol. 48, No. 1, pp. 58–62, 2015

0736-4679/$ - see front matter

ELSEVIER

http://dx.doi.org/10.1016/j.jemermed.2014.07.041

Selected Topics: Neurological Emergencies

EYE ON CHILDREN: ACUTE WORK-UP FOR PEDIATRIC HORNER'S SYNDROME. CASE PRESENTATION AND REVIEW OF THE LITERATURE

Joseph A. Cahill, MD* and Joshua Ross, MD, FAAP†

*PGY3 Neurology, University of Wisconsin Hospitals and Clinics, Madison, Wisconsin and †Pediatric Emergency Medicine, Director of Simulation, Emergency Medicine & Pediatrics, University of Wisconsin School of Medicine and Public Health, Madison, Wisconsin
Reprint Address: Joseph A. Cahill, MD, University of Wisconsin Hospitals and Clinics, 600 Highland Ave., Madison, WI 53792

☐ **Abstract—Background: Ptosis and anisocoria in a child may be subtle indications of occult pathology, and making the observation acutely in the emergency department (ED) is important in guiding patient management and treatment. Emergency physicians must evaluate patients to exclude serious or life-threatening emergencies and ensure correct disposition of patients. Horner syndrome in children may be considered congenital or acquired and may be from benign or malignant causes. When an isolated, acquired Horner syndrome is suspected in a pediatric patient, physical examination of the neck and abdomen for masses, as well as spot urine catecholamines, vanillylmandelic acid and homovanillic acid, and varying degrees of imaging are recommended as part of the initial evaluation. These evaluations may be performed in the ED or may require hospitalization, depending on the suspected anatomical localization and diagnostic considerations. Case Report: A 21-month-old, normally developed girl presented to the University Hospital ED with a 2-h history of right-sided eyelid drooping in the setting of a febrile illness. An eventual diagnosis of Horner syndrome from cervical lymph node compression was made on the basis of history, examination, and imaging findings. Why Should an Emergency Physician Be Aware of This?: ED evaluations of pediatric patients differ from adults. Evaluation and work-up of Horner syndrome in children can be challenging and can require varying degrees of assessment and evaluation, depending on the diagnostic considerations. This article will address the common pathologies responsible for isolated pediatric Horner syndrome and the recommended ED evaluation. © 2015 Elsevier Inc.**

☐ **Keywords—neurology; ophthalmology; Horner syndrome; neuroblastoma**

INTRODUCTION

Eye findings such as ptosis and anisocoria in a child may be subtle indications of occult pathology, and making the observation acutely in the emergency department (ED) is important in guiding patient management and treatment. Emergency physicians must evaluate patients to exclude serious or life-threatening emergencies and ensure correct disposition of patients. Horner syndrome must be among the differential diagnoses in the setting of ptosis with or without anisocoria. How does one make the distinction and what work-up should be done in the ED?

This article will address the common pathologies responsible for isolated pediatric Horner syndrome and the recommended ED evaluation.

CASE REPORT

A 21-month-old, normally developed girl presented to the University Hospital ED with a 2-h history of right-sided eyelid drooping. The mother reported a 3-day history of fever in the setting of an ongoing upper respiratory infection. On examination, she had a right-sided ptosis and mild anisocoria, with the right pupil noted to be roughly 3 mm and the left, 4 mm. Both pupils were briskly

RECEIVED: 19 February 2014; FINAL SUBMISSION RECEIVED: 21 June 2014;
ACCEPTED: 1 July 2014

reactive to light and the asymmetry was made more apparent in dim light. She had no other focal neurological deficits, including ophthalmoparesis, hemiparesis, or mental status changes. There was no history of previous similar episodes. She was admitted for further work-up; a magnetic resonance imaging (MRI) study of the head and neck was performed, which demonstrated diffuse cervical lymphadenopathy. Urine catecholamine testing was not performed. A diagnosis of Horner syndrome from cervical lymph node compression was made. She was eventually discharged with outpatient follow-up in the pediatric clinic. Her ptosis and anisocoria eventually resolved on its own 3–4 days later.

DISCUSSION

Examination Considerations

When an adult or pediatric patient presents with new-onset ptosis to the ED, what are the main considerations? How should one focus the rest of the examination? Obviously, if injury to the eye or eyelid is reported or periocular rash is apparent, the cause is likely nonneurological. If a neurological cause is suspected, are other focal neurological deficits present? Specifically, are there limitations with eye movements or is anisocoria present? Are there other cranial nerve deficits? Also, is the ptosis fatigable (does it vary throughout the course of the day), especially if the patient is reading or focusing? If fatigability or other focal neurological findings are present, then the pattern of signs and symptoms should guide the examination and work-up. Isolated ptosis with anisocoria makes the differential probabilities narrow (Figure 1), with the most likely diagnosis being a third nerve palsy or Horner syndrome.

Ptosis may be subtle and can be assessed by measuring the mid-pupillary distance, or distance from the upper eyelid to the mid pupil. If a patient presents acutely with eyelid ptosis, the first consideration must be, is anisocoria present? If a patient has unequal pupils, which one is abnormal? Eyelid ptosis should point to the abnormal pupil regardless of whether it is the bigger or smaller of the two. Assessing the pupils in both bright light and darkness can help determine which pupil is abnormal. An abnormally constricted pupil should not dilate appropriately in a darkened room, and an abnormally dilated pupil should not constrict in bright light. Either situation will make the difference in size of the pupils greater (due to the normal functioning one), causing a more apparent anisocoria. If the difference between the sizes of the pupils remains the same regardless of either situation, then we might consider the anisocoria physiologic in most cases (with rare exceptions). Apraclonidine or pilocarpine drops can be useful in determining whether the dilated or constricted pupil is physiologic or pathologic, but are rarely used in the ED setting and will not be discussed here. The evaluation becomes simpler if ptosis is present. When both anisocoria and eyelid ptosis are present, one must consider two main diagnostic possibilities: a third nerve palsy (parasympathetic pathway) or a Horner syndrome (sympathetic pathway). Each pathway acts as an antagonist to the other, and disruption of one will cause unopposed stimulation of the other. In most cases, a third nerve palsy will typically have disconjugate

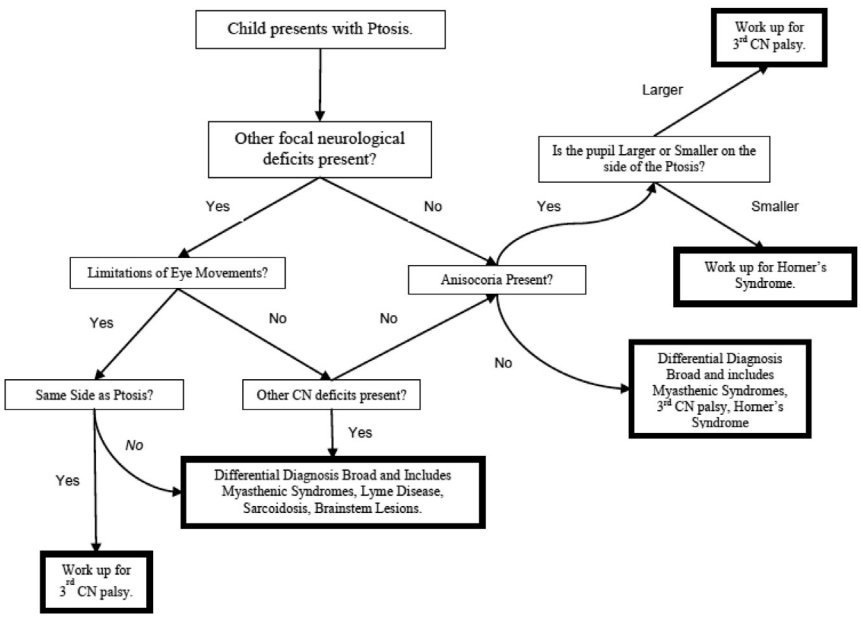

Figure 1. Pediatric ptosis evaluation flow chart. CN = central nerve.