Clinical Images

Horner's, Heterochromia, and Harlequins

Avital Y. O'Glasser, MD, FACP¹ and Kresimira M. Milas, MD, FACS²

¹Division of Hospital Medicine, Department of Medicine, Oregon Health & Science University, Portland, OR, USA; ²Division of Surgical Oncology, Department of Surgery, Oregon Health & Science University, Portland, OR, USA.

KEYWORDS: Horner's syndrome; Harlequin syndrome; Pediatric malignancy; Adult survivors of childhood cancer; Heterochromia.

J Gen Intern Med 31(1):137

DOI: 10.1007/s11606-015-3293-7

© Society of General Internal Medicine 2015

Fig. 1 The patient's affected left eye was different in color, with subtle pupil asymmetry and ptosis.

A 37-year-old woman presented with papillary thyroid cancer as a complication of prior chest and neck radiation to treat a posterior mediastinal ganglioneuroblastoma, for which the patient underwent surgical resection as a child. The ganglioneuroblastoma obliterated the left second intercostal space and extended into the neck along the sympathetic chain, which was transected during resection, producing postoperative Horner's syndrome. Additional late-onset complications of radiation treatment included mild aortic stenosis.

Physical exam revealed heterochromia iridis (Fig. 1), ptosis, and anisocoria. The patient also noted the absence of left facial and arm flushing with heat exposure, exercise, or consumption of spicy foods.

Horner's syndrome produces the triad of ptosis, miosis, and anhidrosis due to damage to the ipsilateral sympathetic chain. The differential diagnosis is broad but should be driven by

patient-specific factors. Congenital or acquired Horner's syndrome in children can lead to heterochromia iridis.¹⁻⁴ Sympathetic stimulation has a trophic effect on iris melanocytes, and denervation impairs melanin pigmentation, making the affected eye lighter.^{3,5-7} The patient's unilateral anhidrosis and absence of flushing was consistent with Harlequin syndrome, which is rare but can occur with Horner's syndrome.⁸

This case exemplifies multiple complications of pediatric malignancy treatment and highlights the importance of cancer survivorship clinics, in which this patient was enrolled.

Conflict of Interest: The authors declare that they do not have a conflict of interest.

Corresponding Author: Avital Y. O'Glasser, MD, FACP; Division of Hospital Medicine, Department of Medicine, Oregon Health & Science University, 3181 SW Sam Jackson Park Road, BTE 119, Portland, OR 97239, USA (e-mail: oglassea@ohsu.edu).

REFERENCES

1. Ropper AH, Brown RH. 14: disorders of ocular movement and pupillary function. In: Ropper AH, Brown RH, eds. *Adams and Victor's Principles of Neurology*. 8th ed. New York: McGraw-Hill Professional; 2005:222-45.
2. Renard D, Jeanjean L, Labauge P. Heterochromia Iridis in congenital Horner's syndrome. *Eur Neurol*. 2010;63(4):253.
3. Sayed AK, Miller BA, Lack EE, Sallan SE, Levey RH. Heterochromia iridis and Horner's syndrome due to paravertebral neurilemmoma. *J Surg Oncol*. 1983;22(1):15-6.
4. McRae D Jr, Shaw A. Ganglioneuroma, heterochromia iridis, and Horner's syndrome. *J Pediatr Surg*. 1979;14(5):612-4.
5. Disenhouse MC, Palay DA, Newman NJ, To K, Albert DM. Acquired heterochromia with Horner syndrome in two adults. *Ophthalmology*. 1992;99(12):1815-7.
6. Imesch PD, Wallow IH, Albert DM. The color of the human eye: a review of morphologic correlates and some conditions that affect iridial pigmentation. *Surv Ophthalmol*. 1997;41:S117-S123.
7. Gladstone RM. Development and significance of heterochromia of the iris. *Ann Neurol*. 1969;21:184-191.
8. Guilloton L, Demarquay G, Quesnel L, De Charry F, Drouet A, Zagnoli F. Dysautonomic syndrome of the face with Harlequin sign and syndrome: Three new cases and a review of the literature. *Rev Neurol (Paris)*. 2013;169(11):884-91.

Received December 4, 2014

Revised January 8, 2015

Accepted March 11, 2015

Published online April 2, 2015