An 8-month-old Baby Girl with Slight Drooping of Left Upper Eyelid

Abstract
An 8-month-old baby girl, who accompanied her sister with an eye problem, was incidentally noticed to have smaller left eye compared to the right. The mother said that it had been present for 2 months. The child showed no detectable focal neurological deficits. Her vision was age appropriate in both eyes. A slight left-sided ptosis was present. Her eye movements were full in both eyes and there was no evidence of strabismus. Bilateral fundus examination was normal. The child had anisocoria (left pupil being smaller than the right one) and left hemifacial anhidrosis, which prompted the diagnosis of Horner’s syndrome. Magnetic resonance imaging of her chest showed a mass lesion in the apical region of the left lung which was later excised and found to be a neuroblastoma. This emphasizes the importance of having an eye on the siblings of children who come to the hospital.

Keywords: Acquired Horner’s syndrome in infants, neuroblastoma, Pupil

Case Report
An 8-month-old baby girl accompanied her parents who had brought her older sister to the eye clinic for treatment of squint in January 2015 at Vijayawada, in the Indian state of Andhra Pradesh. On a casual look, the ophthalmologist noted that the baby’s left eye was smaller than the right eye. The mother indicated that it had been so in the previous 2 months and that they had not noticed any problem with child’s vision. The mother also revealed a history of lack of sweating on the left side of her face. There was no history of any injury or surgery involving any part of the body. On systemic examination, the child did not have any focal neurological deficits. Her visual acuity was eight cycles per degree in both eyes as measured by Teller Acuity Cards. There was mild ptosis in left eye [Figure 1]. She did not have any squint and her eye movements were full in both eyes. She did not have iris heterochromia (iris in affected eye being more lightly pigmented than the other eye). Fundus examination in both eyes did not reveal any abnormality. As seen in Figure 2, the child had a smaller pupil in the left eye (about 2 mm), compared to the right eye (about 2.5 mm) and this anisocoria was more pronounced in the dark than in light (right pupil about 4 mm, left pupil 2 mm as seen in Figure 3). The left pupil did not dilate as readily in the dark, as did the right pupil (“dilatation lag”). Both pupils reacted well to direct and consensual light reflex. Going by these clinical findings and history, a diagnosis of left acquired Horner’s syndrome was made. Magnetic resonance imaging (MRI) of her chest showed a mass lesion measuring 25 mm × 15 mm × 27 mm encasing the vertebral artery, and involving a neurogenic tumor, in the apical region of the left lung, within the left paraspinous superior sulcus [Figure 4]. The child was later referred...
to a surgical oncologist who performed an excision biopsy of the lesion. Histopathology revealed a poorly differentiated neuroblastoma and on immunohistochemistry, a component of ganglioneuroma was also found. A thorough workup was done to look for metastasis, including bone marrow biopsy, whole body positive emission tomography (PET) scan, and iodine 123 (I123) metaiodobenzylguanidine (MIBG) scan, which were all negative. The pediatric oncologist decided not to give chemotherapy or radiotherapy to the child and recommended observation with chest X-ray every 3 months for one year and thereafter every 6 months. The reasons for this approach were the young age at presentation (8 months), the child being negative for N-Myc gene amplification, and the fact that the tumor could be completely excised. Over a period of 6 years follow-up, the child is healthy without any recurrence of the tumor.

**Discussion**

Johann Friedrich Horner, a Swiss ophthalmologist, described the classic clinical trial of signs seen in Horner’s syndrome (ptosis, miosis, and anhidrosis) in 1869 in a 40-year-old woman.[1] It is caused by a lesion anywhere along the sympathetic pathway to the eye, right from hypothalamus, brain stem, and cervical spinal cord (first-order neuron), mediastinum (second-order neuron), and neck or intracranial cavity (third-order neuron). Fibers meant for sweat glands of one side of the face accompany this pathway till the superior cervical ganglion (i.e., second-order neuron). At bifurcation of common carotid artery, these fibers leave the ocular sympathetic pathway and travel to the face along the external carotid artery. Hence, the absence of sweating on one side of face (facial anhidrosis) is a useful sign to localize the site of lesion in Horner’s syndrome to second-order neuron, as in this case.[2]

This case demonstrates that potentially life-threatening conditions like neuroblastoma can sometimes present with very subtle ophthalmic signs in an otherwise asymptomatic child. As in this case, if a child presents with acquired ptosis, the treating physician should first look for anisocoria. If anisocoria is present, the next task should be to assess which pupil is abnormal (smaller or the larger one), by examining pupils in bright light and in the dark. A small pupil that fails to dilate in a darkened room, and a large pupil

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Figure 1: Clinical picture of the child showing mild ptosis in left eye

Figure 2: Mild anisocoria in light (diameter of right pupil about 2.5 mm and left pupil 2 mm)

Figure 3: Anisocoria increasing in the dark since left pupil fails to dilate (diameter of right pupil about 4 mm and left pupil 2 mm)

Figure 4: MRI chest showing a mass lesion in apical region of left lung
that fails to constrict in bright light, is the abnormal pupil. Another practical clue is, usually, the pupil in the eye with ptosis is the abnormal pupil. Then one should check eye movements carefully. Ptosis with a dilated pupil and limited eye movements (all except abduction and intorsion) points towards oculomotor nerve palsy. Ptosis with a smaller pupil and normal eye movements suggests diagnosis of Horner’s syndrome.[1] One specific history that prompted diagnosis of Horner’s syndrome in this case was lack of sweating on one side of face, that is, “hemifacial anhidrosis.” It can involve the entire half of the face or a small patch on the forehead depending on the lesion.[3] It is usually difficult to demonstrate this clinical sign in the clinic. Apart from prompting diagnosis of Horner’s syndrome, unilateral facial anhidrosis also localizes the site of lesion to second-order neuron of sympathetic innervation to the eye.[2]

In children, the causes of Horner’s syndrome can be subdivided into congenital and acquired ones. Important congenital causes include birth trauma (brachial plexus injury), neoplasm, and carotid abnormalities. Acquired cases can be due to any head, neck, and chest surgery, neoplasm, or infection affecting the oculosympathetic pathway.[4] The history often reveals an obvious cause, such as trauma or neck/chest surgery, or that Horner’s syndrome had been present since many years and therefore may not require an extensive investigation. The most common neoplasm presenting with Horner’s syndrome in children is neuroblastoma. It is estimated that about 1 per 10 children of Horner’s syndrome, may have neuroblastoma[3] and 1.3% of neuroblastomas may present with only Horner’s syndrome.[5] Horner’s syndrome in a child of any age without a clear history of trauma or neck/chest surgery requires MRI scanning (with and without contrast), of the brain, neck, and chest.

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Conflicts of interest
There are no conflicts of interest.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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