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Horner's Syndrome in an Infant with Complicated Pneumonia

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Horner's syndrome consists of the classic triad of unilateral miosis, eyelid ptosis, and facial anhydrosis. It occurs as the result of a lesion anywhere along the oculosympathetic pathway. Horner's syndrome as an acquired condition is very uncommon in pediatric neuro-ophthalmology practice and is rarely seen in the pediatric intensive care unit. We present the case of a girl who developed Horner's syndrome during her hospitalization in the PICU for the management of necrotizing pneumonia.

PATIENT DESCRIPTION

The parents gave their informed consent for this case presentation. A 7 month old girl was admitted with the diagnosis of right-sided pneumonia. Chest X-ray showed a large pleural effusion with pneumothorax and partial lung collapse. Computed tomography scan confirmed the diagnosis and a CT-guided thoracostomy tube was inserted. She developed severe hypoxic respiratory failure and had to be intubated and ventilated. During the next 2 days, three additional thoracostomy tubes were inserted due to recurrent tension pneumothoraces. A percutaneous right internal jugular vein catheter was placed and the catheter position was confirmed using standard chest X-ray. Cultures from the pleural fluid grew *Staphylococcus aureus*. On the fifth day of admission, mild anisocoria (1–2 mm) was observed and pediatric neurology and ophthalmology specialists were consulted. On the tenth day of admission the chest drains were removed. The child was extubated with no complications. She was weaned off heavy sedation and slowly returned to normal activity.

Three days after the removal of the drains, a right upper lid ptosis together with the anisocoria were noticed. Ophthalmic examination revealed a right upper eyelid ptosis with slight elevation of the lower lid and anisocoria (the left normal pupil was 3 mm and the right pupil 2 mm in diameter) with increased anisocoria in darkness and mild anhydrosis on the right side of the face. Ocular movements were within normal range. Both pupils were reactive to light. The posterior segment was unremarkable in both eyes. Neurological examination of the cranial nerves and the upper limbs were normal. Instillation of freshly prepared 4% cocaine solution showed the left (normal) pupil dilate to 6 mm and the right pupil to only 4 mm [Figure A], confirming right-sided Horner's syndrome.

At the 6 month follow-up visit in the ophthalmology outpatient clinic, a slight ptosis in the right eye could still be noticed. The anisocoria and anhydrosis were completely resolved.

COMMENT

We present a case of an infant who developed Horner's syndrome while being treated for complicated pneumonia in the PICU.

Horner syndrome was first described in animals (in 1852) by the French physiologist Claude Bernard. In 1869, a Swiss ophthalmologist, Johann Friedrich Horner, was the first to give a complete description of this syndrome in humans. The oculosympathetic palsy called Horner's syndrome appears when the sympathetic innervation of the eye is interrupted by an injury to the neuronal pathway connecting the hypothalamus through the spinal cord to the eye. Mild-to-moderate upper lid ptosis, slight elevation of the lower lid, and pupilary miosis are found in all patients regardless of the severity of the injury. Depending on the level of the lesion, impaired flushing and sweating may be found ipsilaterally. Iris heterochromia (with the affected eye being hypopig-

[A] Right-sided Horner's syndrome. Ptosis: the right upper and lower eyelids are slightly closed. Anisocoria: the left (intact) pupil is 6 mm and the right pupil is 4 mm

PICU = pediatric intensive care unit

mented) is seen in congenital Horner's syndrome [1].

The diagnosis is usually made clinically together with provocative tests. Applying cocaine solution to both eyes will cause the healthy pupil to dilate maximally, while the affected pupil will dilate poorly. Use of 1% hydroxyamphetamine eye-drop solution helps to localize the lesion between the second and third-order neuron lesions [1]. In this case, the etiology was clearly acquired and occurred during the course of the PICU admission.

A literature review revealed that the etiology of pediatric Horner's syndrome varies. Generally it is an uncommon finding in pediatrics and is rare in the PICU environment. Two pediatric case reports described an association between a thoracostomy tube tip placed at the apex of the lung and development of Horner's syndrome [2]. Guccione et al. [3] reported a case of Horner's syndrome in association with central venous line placement into the internal jugular vein in a child [3]. The mechanism by which internal jugular vein cannulation may cause this disorder could be a complication during the procedure, such as puncture of the vessel, or a direct injury to the neuronal pathway during the line placement attempt. It is less clear when the procedure was done without complications and there was no evidence of vascular injury or hematoma. Intrapleural infections such as empyema have been described in association with Horner's syndrome [4]. Increased intrapleural pressure, as in tension pneumothorax, can irritate the stellate ganglion, while relief of the pressure reversed the Horner's syndrome symptoms. Overall, most investigators suggest that irritation of the second neuron pathway and stellate ganglion is the most common acquired etiology and is usually multifactorial.

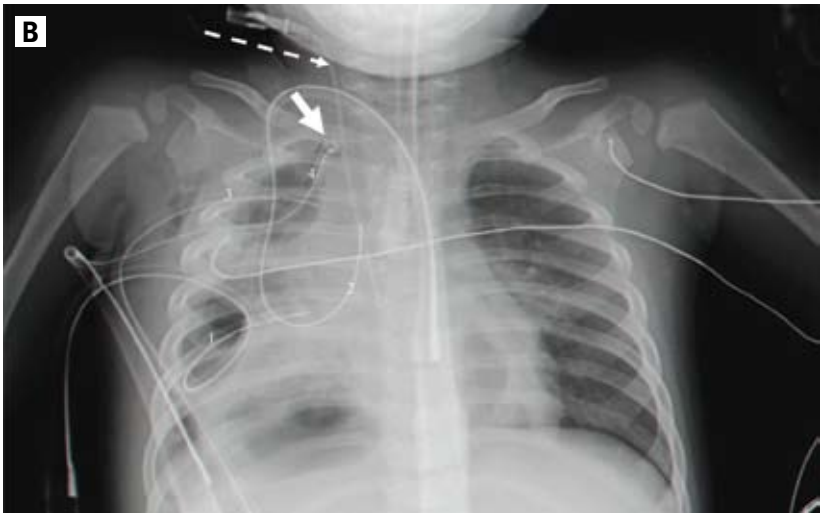
Our patient had several conditions that increased her risk of developing an injury to the oculosympathetic pathway. She was admitted with a complicated necrotizing pneumonia and developed thoracic empyema. Her medical condition necessitated aggressive ventilation using high pressure and she developed pneumothoraces. During her treatment, she had several episodes when pleural tubes were inserted. During her critical illness, an internal jugular vein catheter was placed.

The first thoracic (or stellate) ganglion lies in close proximity to the parietal pleura of the lung apex and only a thin layer of connective tissue, the endothoracic fascia, separates these two structures. The great upper body blood vessels, including the internal jugular vein, lie in close proximity to the ganglion. Any process that involved this anatomic space could have caused an injury to the ganglion: inflammation due to infection and empyema, baro/volume trauma from pneumothorax, a direct injury to the nerve caused by the tip of the chest tube placed at the apex of the thorax by itself or by "trapping" the ganglion between the jugular catheter and the chest tube, dissection of any of the great head blood vessels, or a hematoma from vascular manipulation during the insertion; all or some could be considered as possible mechanisms. Our patient's chest X-ray shows the location of the foreign hardware that was inserted [Figure B]. The thick white arrow points to the tip of the chest tube (marked as number 4), which was positioned at the apex of the right lung and its proximity of the right jugular venous catheter (marked with the dashed arrow). It is clearly seen that both are adjacent to the anatomic location of the stellate ganglion.

Although the natural course of Horner's syndrome is not predictable, there is some evidence that the time length of the injuring process or pressure applied on the second-order neuron might be related to outcome. Kaya and team [5] found that in adults with Horner's syndrome secondary to chest tube malposition, early removal of the apical chest tube improved the chance of recovery.

Diagnosing Horner's syndrome in critically ill pediatric patients is difficult unless one suspects it. These children are usually heavily sedated and often chemically paralyzed, which complicates the ability to perform a thorough neurological examination. By presenting this case, we hope to increase awareness regarding the syndrome and the risk of it developing in such patients.

[B] Chest radiograph of our patient during her acute illness shows a right-sided consolidation with diffuse air space disease and four chest drains marked with white numbers 1-4. The dashed arrow points to the right internal jugular vein catheter. The thick white arrow points to the location of the tip of the chest tube (#4) and the internal jugular catheter, both adjacent to the anatomic location of the sympathetic chain and the stellate ganglion.



Early recognition and prompt removal of possible injurious iatrogenic hardware might prevent future permanent disability, which may persist long after the fact is forgotten that the child's life was saved in the PICU. This may also have medico-legal implications for the medical team.

In conclusion, we describe a rare case of Horner's syndrome developing during hospitalization in the PICU. We stress the importance of recognizing this entity. Prompt diagnosis and immediate repositioning of chest drains may prevent irreversible iatrogenic damage. We recommend daily evaluation of pupillary size and presence of ptosis in patients at risk of developing Horner's syndrome (such as the tip of the chest tube at the apex of the lung and its proximity to the internal jugular catheter) in order to prevent missing the correct diagnosis.

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