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CASE STUDY

THE PALPEBRAL FISSURE NARROWING LEADING TO THE NEUROBLASTOMA DIAGNOSIS – A CASE REPORT

ZWĘŻENIE SZPARY POWIEKOWEJ JAKO PIERWSZY OBJAW NEUROBLASTOMA

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ABSTRACT

Introduction

Horner's syndrome manifests itself by anisocoria, one-sided ptosis, anhidrosis, and enophthalmos. It is caused by an interruption of the oculosympathetic tract. Among pediatric patients, the most common etiology is trauma.

Aim

The aim of this paper was to present a case report of an infant with Horner's syndrome who experienced a fall and had a surgery; however, finally was diagnosed with the neuroblastoma.

Material, methods, results

A 3-month-old patient presented with left-sided ptosis. The palpebral fissure narrowing occurred abruptly in the 4th week after birth. After phenylephrine administration the left eyelid lifted. Lack of the pupils' reaction after the 0.1% adrenaline administration, suggested the diagnosis of the preganglionic Horner's syndrome. Moreover, the left pupil miosis was observed. In the age of 1 month, the operation of the pyloric stenosis was performed. The occurrence of drooping eyelid was connected with the surgery or incident of a fall reported by the mother of the patient. The head ultrasound and thoracic RTG did not show any pathologies. Two weeks after ophthalmologic consultation magnetic resonance imaging was performed and revealed a 1.6 cm mass adjacent to the left subclavian artery. The biopsy of the tumor confirmed the diagnosis of the neuroblastoma. The chemotherapy and the surgical removal of the tumor were performed. The child is in complete remission of neuroblastoma and has been under permanent oncologic care since 3 years.

Conclusion

This case report shows that Horner's syndrome requires vigilance and should lead to cervical and brain MRI examination in children because of the possible neuroblastoma incidence.

Key words: Horner's syndrome, ptosis, miosis, anisocoria

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STRESZCZENIE

Wstęp

Do objawów zespołu Hornera należą anizokoria, jednostronna ptoza, zapadnięcie gałki ocznej i zmniejszona potliwość. Jest on spowodowany uszkodzeniem włókien współczulnych unerwiających oko. Najczęstszą przyczyną zespołu Hornera w grupie pacjentów pediatrycznych jest uraz.

Cel

Celem tej pracy jest przedstawienie przypadku niemowlęcia z zespołem Hornera, które przebyło w pierwszym miesiącu życia upadek i operację, jednak ostateczną przyczyną zaobserwowanych objawów okazał się nerwiak zarodkowy.

Materiały i metody, wyniki

Do Kliniki Okulistycznej Szpitala Przemienienia Pańskiego w Poznaniu zgłosiła się matka z 3-miesięczną córką z powodu lewostronnego opadania powieki u dziewczynki. Zwężenie lewej szpary powiekowej wystąpiło nagle w 4. tygodniu życia pacjentki i utrzymywało się przez kolejne dwa miesiące. U pacjentki zaobserwowano zwężenie lewej źrenicy. Po przeprowadzeniu badania z podaniem fenylefryny i 0.1% adrenaliny zdiagnozowano przedzwojowy zespół Hornera. W 1. miesiącu życia pacjentka przebyła operację zwężonego odźwiernika. Matka dziewczynki opadanie powieki kojarzyła z upadkiem z kanapy przez pacjentkę. USG głowy i RTG klatki piersiowej nie wykazały żadnych patologii. Dopiero badanie rezonansu magnetycznego pozwoliło uwidocznić zmianę o średnicy 1.6 cm w okolicy lewego obojczyka. W wyniku biopsji potwierdzono podejrzenie neuroblastoma wysunięte przez radiologów. Dziewczynka była leczona chemioterapią i operacyjnie. Aktualnie ma 3 lata, jest pod stałą opieką onkologa, pozostaje w pełnej remisji choroby.

Wnioski

Przypadek tej pacjentki pokazuje, że zespół Hornera wymaga uwagi, a zdiagnozowanie jego przyczyny powinno opierać się na badaniu rezonansu magnetycznego głowy i szyi.

Słowa kluczowe: Zespół Hornera, opadanie powiek, zwężenie źrenic, anisokoria

Introduction

Horner's syndrome is the set of unilateral symptoms due to an interruption of the oculosympathetic tract. The syndrome manifests itself by miosis, ptosis, enophthalmos, and anhidrosis. The incidence of Horner's syndrome among children younger than 19 years old is 1.42 per 100 000 (Smith *et al.* 2010). It is divided into acquired and congenital. In both groups, the trauma is the leading cause of the incidence of the symptoms and constitutes 65% of the cases (Smith *et al.* 2010). However, Horner's syndrome can be the result of a tumor as well, the most often neuroblastoma. Neuroblastoma has an incidence of 27.75 per 100 000 patients younger than 5 years and is the most common extracranial solid malignant tumor in this group of patients (Bernstein *et al.* 2016). The most frequent localization is the abdomen, especially the adrenal medulla. However, back mediastinum, neck and pelvis are also possible localizations of neuroblastoma.

Aim

The aim of this paper was to present a case report of an infant with Horner's syndrome

which was assumed to be connected with trauma or surgery performed previously. However, finally, the patient was diagnosed with neuroblastoma.

Patient, methods and results

A mother of a 3-month-old girl with the left-sided ptosis attended the ophthalmologic department. The palpebral fissure narrowing occurred abruptly in the 4th week after birth. Earlier, the child was consulted with 3 specialists (2 ophthalmologists and a neurologist), and no pathologies were detected. The mother reported a trauma – a fall from the sofa in the first month of life. Moreover, at the age of 1 month, the operation of the pyloric stenosis was performed. During the control visit after the surgery, the thoracic RTG was performed and did not show any pathologies. The girl was born at the 40th week of pregnancy with the birth weight of 3480 g and 10 points in the Apgar Score. There were no complications during labor.

At the ophthalmologic clinic, the full eye examination was performed. The anterior segment of the eye was normal. Moreover, the examination of the eye fundus and ultrasound did not reveal any pathologies. No nystagmus or strabismus were detected. However, after 2.5% phenylephrine administration the left eyelid lifted. Lack of the pupils' reaction to the 0.1% adrenaline administration, suggested the diagnosis of the preganglionic left-sided Horner's syndrome. Moreover, the left pupil narrowing was observed. The distance between the pupil and the edge of the upper eyelid was 4 mm in the right eye, and 2mm in the left eye. Motility of the eyes and pupils' light reactions were normal.

The girl was admitted to the clinic, and the MRI was performed and revealed a 1.6 cm mass adjacent to the left subclavian artery and surrounding the proximal part of the left common carotid artery. A reinforcement of the mass was observed after an administration of the contrast. The change was adjacent to the left lung, and also to the C7/Th1 vertebral body. The suspicion of the neuroblastoma was suggested. The scintigraphy MIBG showed one focus of gathered radiofarmaceutic close to the left clavicle.

The biopsy of the tumor revealed small-cell invasive mass with Ki-67 index value of 40%. The histological examination confirmed the diagnosis of the neuroblastoma. The bone marrow biopsy denied MYCN gene amplification. However, it showed cell-rich tissue without CD56 cells of neuroblastoma morphology (2%) and the growth in the lymphocytes number.

Because of the localization and size of the tumor, the surgery of the neuroblastoma was postponed. The chemotherapy was administered, and then the surgery of the tumor was performed. The child in in complete remission of neuroblastoma and has been under permanent oncologic care since 3 years.

Discussion

Congenital Horner's syndrome is more frequent than acquired (55% vs. 45%), and the most common cause is a birth trauma (Smith *et al.* 2010). However, in this case, the symptoms were observed 4 weeks after birth, and there were no problems during or directly after the labor. So, the girl had acquired Horner syndrome. Her mother and the doctors initially connected the incidence of syndrome with the surgery and the fall which the patient experienced in the first month of life. The most frequent cause of acquired Horner's syndrome is the trauma, so this correlation was understandable. Nevertheless, in other analyses, the incidence of the neuroblastoma as a cause of Horner's syndrome fluctuates from 0% to 42% (Smith *et al.* 2010). This variability has the consequences in changing recommendations of Horner's syndrome causes diagnosis.

Acquired Horner's syndrome is divided into 3 groups: central preganglionic lesion, preganglionic lesion of the second neuron of the oculosympathetic tract, caused by neck trauma, neuroblastoma or another tumor, and postganglionic lesions (Lambert *et al.* 2017). The patient presented in this paper

was diagnosed with preganglionic Horner syndrome. According to Taylor and Hoyt additional imaging is required in the cases of non-obvious trauma or surgery etiology. The consultation with the neurologist is also significant (Lambert *et al.* 2017).

The diagnosis of neuroblastoma is based on the imaging. However, MRI or CT examination in an infant performed under general anesthesia correlates with significant risk in an infant. The other way to evaluate the risk of neuroblastoma is an examination of the level of urine catecholamine. Nonetheless, there are many cases of diagnosed tumors whereas these parameters are not elevated (Smith *et al.* 2010). Accordingly, the normal level of the urine catecholamine cannot exclude the neuroblastoma.

There are many publications analyzing the incidence of neuroblastoma in patients with Horner syndrome. Published studies indicate that Horner's syndrome diagnosed in the first year of life is connected usually with a benign lesion (George *et al.* 1998). In isolated syndrome the routine CT or MR are unnecessary and should be performed only in cases associated with additional systemic signs (George *et al.* 1998; Shabat et a. 2019) Other research papers imply that Horner's syndrome occurs frequently as a first sign of neuroblastoma presence (Alvi *et al.* 2017), and which has subtle or unspecific manifestation (Olivera *et al.* 2018; Cohen *et al.* 2018).

According to the author's knowledge, there is no publication in the literature presenting a case of an infant with the manifestation of Horner's syndrome and such a history of the risk factors for these symptoms. Despite reported trauma and performed surgery, the careful examination of a child with Horner's syndrome should be extended, in some cases including imaging examinations.

Conclusion

This case report shows that Horner's syndrome requires vigilance and should lead to cervical and brain MRI examination because of the neuroblastoma suspicion. It is difficult to define the guidelines for the diagnostic process of Horner's syndrome in children.

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