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Case Report

Acute Strabismus as the First Sign of Optic Nerve Glioma - Diagnostic Challenge

Biljana Trajkovic[1]* and Ivana Todorovic[2]

[1]KBC Zvezdara Eye Clinic, "Dr. Ivan Stankovic", Faculty of Medical Science, Kosovska Mirtovica, Serbia

[2]KBC Zvezdara Eye Clinic, "Dr. Ivan Stankovic", Belgrade, Serbia

***Corresponding Author:** Biljana Trajkovic, KBC Zvezdara Eye Clinic, "Dr. Ivan Stankovic", Faculty of Medical Science, Kosovska Mirtovica, Serbia.

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Abstract

Introduction: Gliomas of the optic pathway are benign tumors of the CNS that primarily affect children. Optic gliomas are rare. The cause of optic gliomas is unknown. It occurs as part of neurofibromatosis. Glioma itself can affect any part of the optical system.

Method: Presentation of a 18 months old girl with acute strabismus as the first sing of optic nerve glioma. Detected by ultrasound examination and confirmed by magnetic resonance.

Case Report: Parents bring a small child-girl, 18 months old, for examination.

Due to the acute outward turning of the right eye, in the last month. Take an anamnestic.

Receives information about "café au lait" spots on the body (spots the color of white coffee) with suspicion to neurofibromatosis type. On the fundus of the right eye edema of the optic nerve, the left optic nerve fundus is temporally pale. On ultrasound a large orbital mass is detected in the region of the optic nerve of the right eye.

Conclusion: The aim is to diagnose and treat all children's ophthalmological diseases an interdisciplinary approach that leads to timely diagnosis and visual preservation functions.

Keywords: Glioma; Strabismus; Neurofibromatosis; Optic Nerve; Ultrasound

Introduction

Gliomas of the optic pathway are benign tumors of the CNS that primarily affect children [1]. Optic gliomas are rare. The cause of optic gliomas is unknown. Most optic gliomas are slow-growing and noncancerous and occur in children, almost always before age 20. Most cases are diagnosed by 5 years of age.

There is a strong association between optic glioma and neurofibromatosis type 1. The location of these tumors makes surgical treatment impossible without visible consequences.

Treatment of patients with optic pathway glioma requires an interprofessional team.

Symptoms of optic glioma include:

- Vision loss,
- Double vision,
- Crossed eyes, and
- A loss of color vision.

Diagnosis may involve a physical exam and imaging tests such as an MRI or a CT scan. Other tests that may be performed include

an electroretinogram, a visual field test, blood tests, and genetic testing [2].

Materials and Methods

We determined the existence of glioma by ultrasound examination.

Results and Discussion

An 18-month-old child from another controlled pregnancy, born at term, comes for examination due to an acute deviation of the right eye in the field (exotropia),since a month ago.

Objective finding

Child follows small objects and light. In primary position right eye in divergence, with protrusion of eye ball of 1mm (hertel) left eye ortho. Relative afferent pupillary defect positive in the right eye.

Examination of the fundus of the eye with a fundoscopy, we find papile edema of the optic nerve on the right fundus, Left fundus a good finding.

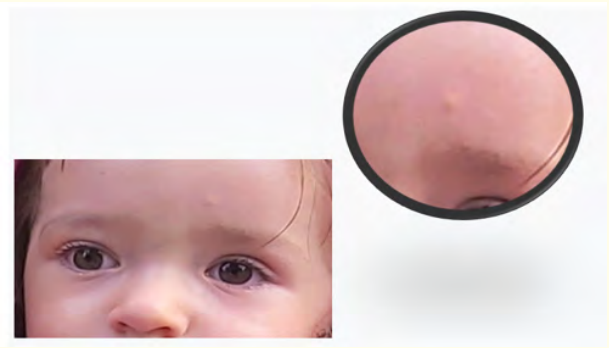

Figure 1

On the child's whole we observe growths that look like a nodus characteristic of neurofibromatosis.

Through a detailed examination of the child, we understand that the entire body is covered with the so-called white coffee stains "café au late".

Is this neurofibromatosis nf 1 and does the child have optic nerve glioma???

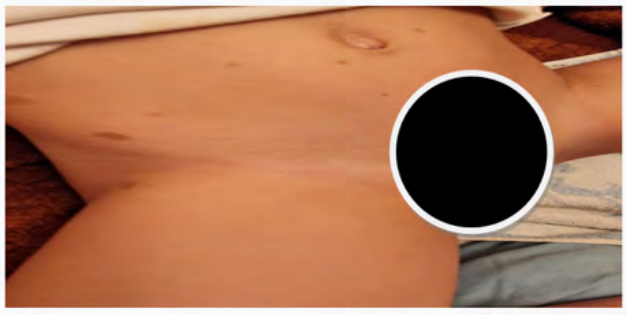

Figure 2

No case of neurofibromatosis has been registered in the family so far.

What to do next!

We applied ultrasound diagnostics, which was immediately available to us.

Figure 3

Using ultrasound, we detected papille edema of the optic nerve of the right eye with a large orbital mass, at that moment the left eye ball was normal.

The diagnostic criteria for NF1 are fulfilled in persons whose parents do not have NF if two conditions are positive:

- Six or more "café au lait" spots - over 5 mm
- Freckles in the axillary or inguinal region
- Two or more neurofibromas
- Optic nerve glioma

- Two or more Lisch nodules on the iris or two or more choroidal abnormalities
- Bone lesions [3].

The child was urgently shoot for a magnetic resonance of the head.

MR findings: Massive infiltration of the right optic nerve from the papilla to the chiasm - morphologically and signally corresponds to a low-grade glioma. Suspicious initial infiltrative change of the left optic nerve intraorbitally.

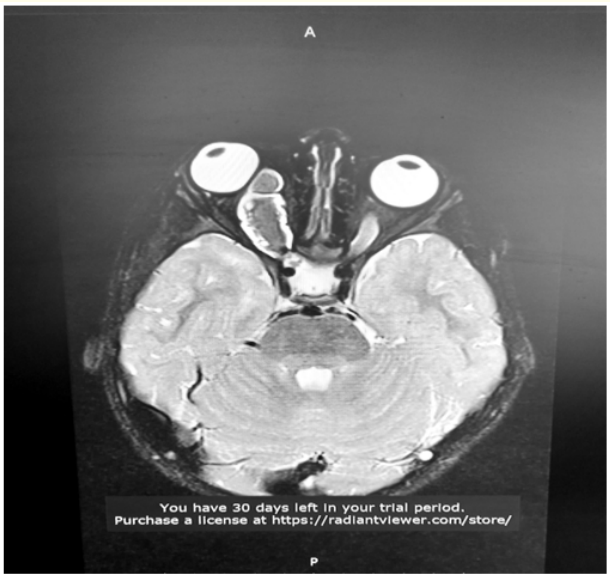

Figure 4

Treatment options for proven glioma are:

- If it is stable - observation clinically and radiologically
- Standard chemotherapy
- Molecularly targeted therapy-inhibitors of the mitogen-activated creatinine kinase pathway (MEK inhibitors selemutinib, refametinib, trametinub and combimetinib)
- Bevacizumab (VEGF)
- Radiotherapy-not for children!!!
- Surgery (orbitotomy approach recommended) [4].

The girl underwent neurosurgery. Craniotomy was performed.

Conclusion

The diagnosis and treatment of all children's ophthalmological diseases aims at an interdisciplinary approach that leads to timely diagnosis and preservation of visual acuity [5].

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Conflict of Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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