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Strabismus and Exophthalmos in Children: Think of Optic Nerve Glioma

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Abstract | **Case Report**

***Introduction*:** Optic nerve gliomas are rare tumors that are mainly observed in children. Exophthalmos and strabismus are the main signs of the disease. Neuroimaging and in particular magnetic resonance imaging is of great help in the diagnosis and follow-up of these tumors. The therapeutic management of these gliomas involves different means: surgical removal, chemotherapy, radiotherapy or abstention under surveillance. The indications must be discussed on a case-by-case basis. We report in this work a case of optic nerve glioma revealed by a strabismus and a unilateral exophthalmos. ***Case Report*:** This is a 3-year-old girl from a non-consanguineous marriage, with no specific pathological ATCD, who presented with divergent strabismus with left exophthalmos evolving for 3 months. The ophthalmologic examination noted a constant wide-angle divergent strabismus on the left eye with moderate exophthalmos on the same side. Corrected visual acuity was reduced to near finger count on the left and 5/10 on the right. Somatic examination found café au lait spots on the back and trunk. Cerebral CT showed a bilateral thickening of the optic nerves, fusiform and regular, more marked on the left, isodense to spontaneous contrast, homogeneously enhancing after injection of PDC. On the left, it is responsible for a downward refoulement of the eyeball with grade 1 exophthalmos. An MRI was ordered revealing a regular well-limited fusiform thickening of the optic nerves in T1 isosignal, intermediate in T2 and FLAIR, in diffusion hypersignal without ADC restriction, intensely and finely heterogeneous enhancing after injection of contrast medium. This thickening is responsible for a grade I left exophthalmos, with widening of the optic canals posteriorly and filling of the intraconical fat evoking a glioma appearance. The child was referred to neurosurgery for management. ***Conclusion*:** The diagnosis of optic nerve glioma in children is rare, with atypical presentations both in terms of discovery and evolution. The possibility of invasion of the optic chiasm as well as bilateralization seriously conditions the visual functional prognosis. The therapeutic management is heavy for a benign tumor which does not involve the vital prognosis and must be multidisciplinary.

Keywords: Optic nerve gliomas, Neuroimaging, diagnosis, tumors, bilateralization.

INTRODUCTION

Optic nerve gliomas are rare tumors that are mainly observed in children. Exophthalmos and strabismus are the main signs of the disease. Neuroimaging and in particular magnetic resonance imaging is of great help in the diagnosis and follow-up of these tumors. The therapeutic management of these gliomas involves different means: surgical removal, chemotherapy, radiotherapy or abstention under surveillance. The indications must be discussed on a case-by-case basis.

We report in this work a case of optic nerve glioma revealed by a strabismus and a unilateral exophthalmos.

CASE REPORT

This is a 3-year-old girl from a non-consanguineous marriage, with no specific pathological ATCD, who presented with divergent strabismus with left exophthalmos evolving for 3 months.

The ophthalmologic examination noted a constant wide-angle divergent strabismus on the left eye with moderate exophthalmos on the same side. Corrected visual acuity was reduced to near finger count on the left and 5/10 on the right. Somatic examination found café au lait spots on the back and trunk.

Cerebral CT showed a bilateral thickening of the optic nerves, fusiform and regular, more marked on the left, isodense to spontaneous contrast, homogeneously enhancing after injection of PDC. On the left, it is responsible for a downward refoulement of the eyeball with grade 1 exophthalmos.

An MRI was ordered revealing a regular well-limited fusiform thickening of the optic nerves in T1 isosignal, intermediate in T2 and FLAIR, in diffusion hypersignal without ADC restriction, intensely and finely heterogeneous enhancing after injection of contrast medium. This thickening is responsible for a grade I left exophthalmos, with widening of the optic canals posteriorly and filling of the intraconical fat evoking a glioma appearance. The child was referred to neurosurgery for management.

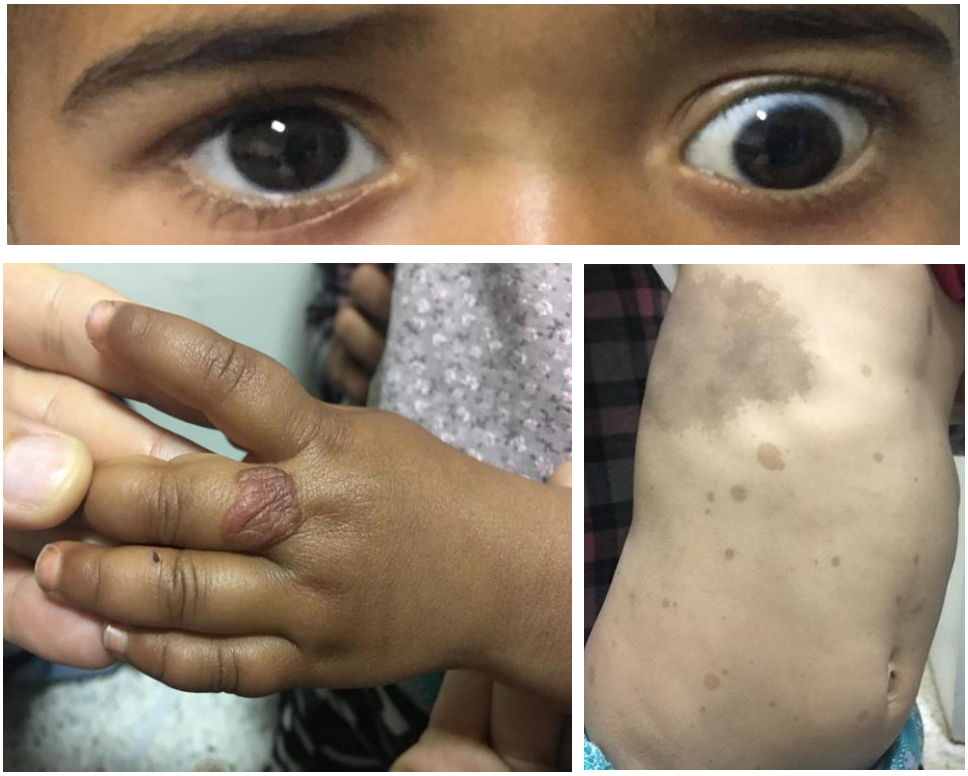

Figure 1: Multiple café au lait spots on the extremities and trunk with left exophthalmos

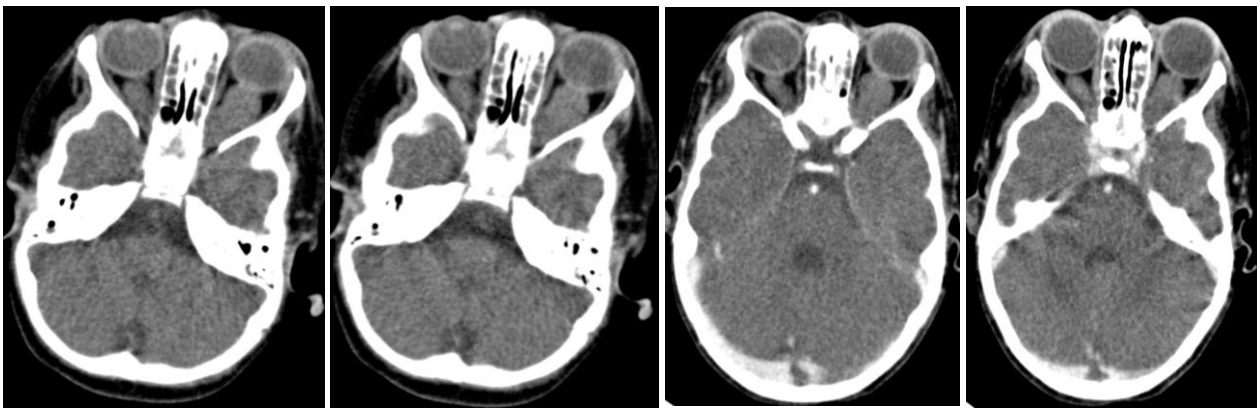

Figure 2: Brain scan with and without PDC injection in axial section: bilateral thickening of the optic nerves, fusiform and regular, more marked on the left, isodense to spontaneous contrast, homogeneously enhancing after PDC injection. It is responsible on the left for a downward refoulement of the eyeball with grade 1 exophthalmos

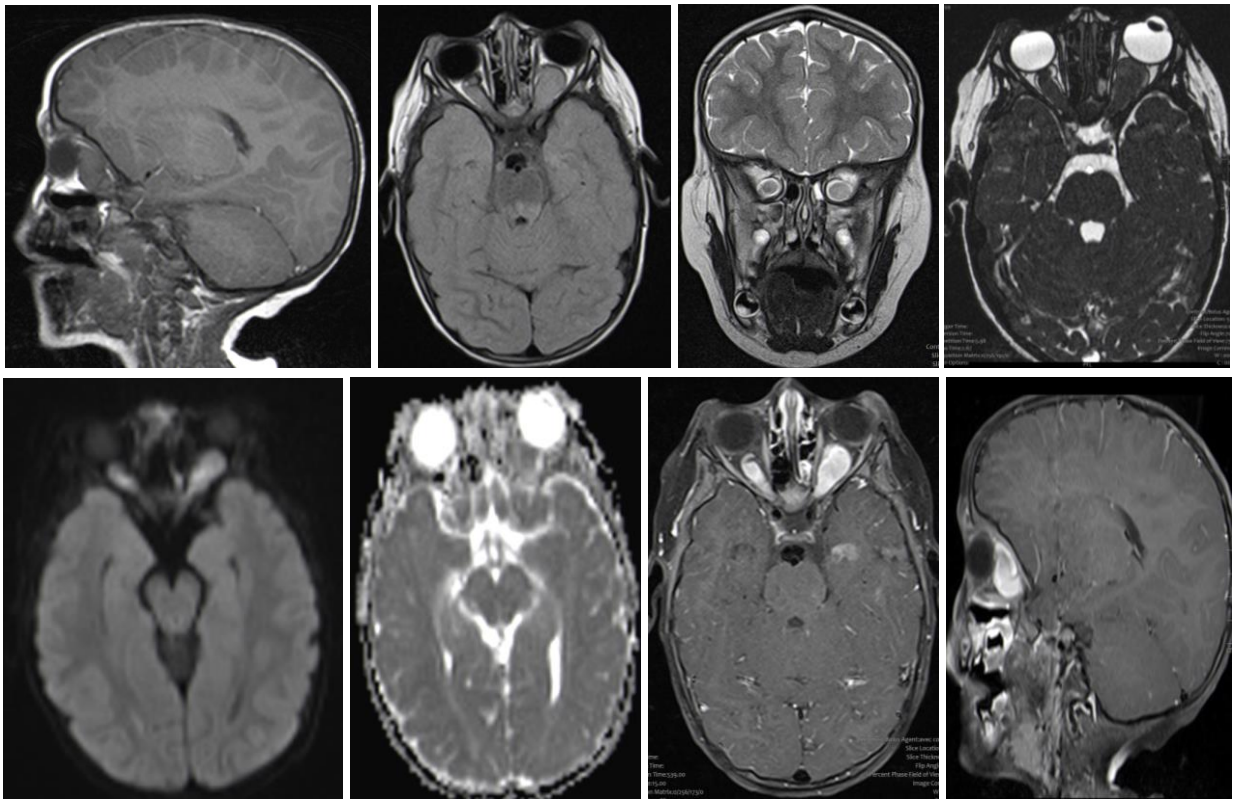

Figure 3: Cerebral MRI in T1 axial, T2 coronal, diffusion and T1 3D sequence after injection of Gadolinium: regular well-limited fusiform thickening of the optic nerves in T1 isosignal, intermediate in T2 and FLAIR, in diffusion hypersignal without ADC restriction, intensely and finely heterogeneously enhancing after injection of contrast medium. This thickening is responsible for a grade I left exophthalmos, with widening of the optic canals posteriorly and filling of the intraconical fat

DISCUSSION

Optic nerve gliomas are rare tumors that occur mainly in children. Bilateral optic nerve involvement is considered a characteristic of these gliomas in the context of neurofribromatosis type I (NF1) as in our patient's case.

Von Recklinghausen disease or neurofibromatosis type I is the most common phacomatosis. It is a genetic disease with autosomal dominant transmission, a multi-systemic pathology but with a mainly neuro-cutaneous tropism.

The diagnosis of the disease is based on 7 cardinal criteria:

- At least 6 café au lait spots (>0.5 before puberty and >1.5 after).
- At least two neurofibromas.
- Axillary or inguinal lentigines.
- Optic nerve glioma.
- Bone lesion: pseudarthrosis, sphenoid dysplasia, thinning of the cortex of the long bones.
- At least two lisch nodules (irian hamartoma).

Exophthalmos and strabismus are often indicative of the disease.

Magnetic resonance imaging is a key examination in the exploration of these gliomas. It allows not only to study the optic nerves but also to search for a possible extension of these gliomas to the orbit, the chiasma and the intracranial structures, particularly in case of association with NF1. It also has the advantage of being less radiating compared to CT scan.

Therapeutic management ranges from simple clinical and radiological monitoring to surgical treatment or chemotherapy or radiotherapy.

The therapeutic indications are discussed on a case-by-case basis and depend on the visual acuity, the degree of exophthalmos and the extension of the tumor. In case of NF1, radiotherapy is formally contraindicated because of the complications it may cause. In our case, therapeutic abstention with clinical and radiological monitoring was recommended.

These controls are done every six months, and to date we have not noted any progression of the lesions. According to Tow *et al.*, the prognosis of gliomas of the optic pathways associated with NF1 seems to be better.

CONCLUSION

The diagnosis of optic nerve glioma in children is rare, with atypical presentations both in terms of discovery and evolution. The possibility of invasion of the optic chiasm as well as bilateralization seriously conditions the visual functional prognosis. The therapeutic management is heavy for a benign tumor which does not involve the vital prognosis and must be multidisciplinary.

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