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Exophytic ganglioglioma of the optic nerve mimicking pilocytic optic pathway glioma

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Introduction: Gangliogliomas arise from ganglion cells and are very rare CNS tumors in kids. They most often occur within the temporal lobe, yet also in the frontal, parietal and occipital lobe, thalamus and third ventricle. Gangliogliomas within the optic pathways are extremely rare with less than a dozen cases reported in the world literature. We report a large exophytic ganglioglioma of the right optic nerve which was treated by surgery alone.

Methods: A three year old girl suffered from deterioration of vision of her right eye to complete blindness. Her left eye had full vision without any impairment of visual field, indicating full function of chiasm. MRI showed a large glioma arising from the right optic nerve and chiasm. The tumour had cystic parts, areas with contrast uptake and hypodense parts, so that a typical pilocytic optic pathway astrocytoma was suspected.

Results: Surgery, using a right fronto-lateral approach, was performed with extensive resection, leaving some of the right optic nerve. Small residual tumour was left inside the right chiasma to preserve its function. The left optic nerve, both optic tracts, pituitary stalk and hypothalamus were preserved. Histopathology reported a ganglioglioma. Postoperative visual function was unchanged as was endocrinology. Since the tumour remnants were stable at 24 months postoperatively, the girl is currently under observation.

Conclusions: Since gangliogliomas of the optic pathways are extremely rare, they are not taken into account as differential diagnosis. Diffuse optico-hypothalamic gliomas are no candidates for radical surgery. However, symptomatic and exophytic ones can be sufficiently treated by surgery alone. Our case demonstrates that a surgical strategy can be successful without additional harm and that surgery can reveal unexpected histologies. Gangliogliomas however do not differ from pilocytic astrocytomas in their treatment approach.