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Optic Pathways Gliomas (OPW) in children. Single institutional experience

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Introduction: Tumors affecting the Optic Chiasm and nerves in children are usually Low Grade Gliomas. In our hospital, selected cases underwent surgical treatment and we wanted to analyse the outcomes.

Patients and Methods: Retrospective study in patients diagnosed with OPW. Several variables were analysed to find out possible prognosis factors and to evaluate the outcomes.

Results: Of 36 cases studied, 21 were female. Mean age at diagnose: 49.4 months (3.1 – 206.7), NF1 associated in 13 cases (36.1%). Main symptoms were visual alterations (61.1% of cases). Hydrocephalus was diagnosed and treated in 14 cases, 43.8%.
After diagnose, progression was observed in 22 cases (61.1%). Twenty five patients were operated (69.4%), achieving 2 Gross Total Resections (GTR), 20 Subtotal resections (STR) and 3 Biopsies. After surgery, 18 patients presented transient neurologic deficits, 4 systemic and 9 local complications. Twenty four patients received Adjuvant Therapy, 66.7%. Histology showed 13 Pylocytic Astrocytomas (52%), 2 LGG “nos” (8%), 8 Fibrillary Astrocytomas (32%).
Mean follow up was 64.2 months (3.8-174.2). At study closure 7 patients were dead due to progression, other 29 were alive (22 with sequels). One patient with GTR was alive with no evidence of disease.
Variables related with worse PFS were: antecedent of NF1 (p=0.018) and use of Adjuvant therapy (p=0.056); worse OS were: Systemic complications (p=0.027), Progression (p=0.02) and age at diagnose (5 of 6 patients younger of one year died).
In our series, mean PFS was 79.5 months. OS at 3, 5 and 10 years were 94%, 85% and 77%.

Conclusions: OPW in children are usually LGG. NF1 was associated to good prognosis for OS but not for PFS. Factors associated with poor prognosis were Age less than one year, Systemic complications and Progression. The benefit of tumor removal was evident but not significant because of the small number of cases.