Optic tract glioma and endocrine disorders- comparison between patients with and without NF1- a single center experience

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ESPE 2021
The 59th Annual Meeting Online
22-26 September 2021

INTRODUCTION

Optic pathway gliomas (OPGs) represent 2-5% of brain tumors in children.

OPGs are classified by the anatomic location and whether they are associated with neurofibromatosis type 1 (NF1).

Children with OPGs face sequelae related to tumor location and treatment modalities, including visual dysfunction, neurologic deficits, and endocrine dysfunction.

AIM

To assess the prevalence of endocrine dysfunctions in patients with OPGs and to compare the outcomes between those with and without NF1.

METHODS & SUBJECTS

A retrospective single center study that included patients diagnosed with OPGs between 1990 and 2020, younger than 18 years at diagnosis, followed at our endocrine clinic.
Data about demographic parameters, presence/ absence of NF1, OPGs treatment related data, growth and puberty parameters and occurrence of endocrine dysfunction were retrieved from their medical records.

- Variables were compared between groups (OPGs with and without NF1)

RESULTS

The cohort included 59 children (29 males), 36 (61%) had NF1, with higher rates of isolated optic nerve involvement (p<0.01)

Percentage of patients with one or more OPG-related endocrine comorbidities by NF1/non NF1

Parameter	Non NF1	NF1	P value
Growth hormone deficiency	3 (13%)	3 (8.3%)	0.66
Growth hormone excess	1 (4%)	1 (2.8%)	0.74
Central hypothyroidism	11 (48%)	1 (2.8%)	<0.01
Precocious puberty	3 (13%)	8 (22%)	0.37
Hypogonadotrophic hypogonadism	5 (22%)	1 (2.8%)	0.02
Diabetes insipidus	6 (26%)	1 (2.8%)	0.01
Hyperlipidemia	8 (35%)	3 (8.3%)	0.01
Primary gonadal failure	2 (8%)	0	0.15

Data is presented as n (%)

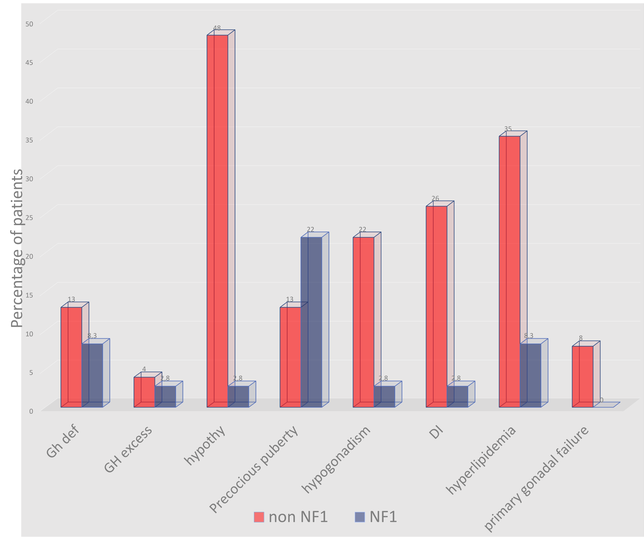
In a multivariate regression analysis, surgery was the independent predictor of endocrine dysfunctions (OR 13.3 [2.23-79.53]; P<0.01)

Comparison of basic characteristics of the cohort between groups (OPGs with & without NF1)

Parameter	Non NF1 (n=23)	NF1 (n=36)	P value
Gender male	11 (48%)	18 (50%)	0.87
Age at diagnosis (years)	1.5 (0.7-7.2)	4.2 (2.5-8.6)	0.04
Age at last visit (years)	14.7 ± 6.3	11.5 ± 5.0	0.03
Tumor location:			<0.01
Optic nerve only	0	11 (30%)	
Optic chiasm ± optic nerve	11 (48%)	17 (47%)	
Hypothalamus or adjacent structures	12 (52%)	8 (22%)	
Years since diagnosis	10.8 ± 6.0	6.6 ± 4.4	<0.01
Treatment modality			
Surgery	15 (65%)	3 (8%)	<0.01
Chemotherapy	19 (83%)	11 (31%)	<0.01
Other therapy	6 (35%)	6 (18%)	0.18
Radiation	3 (14%)	1 (3%)	0.13
Tanner at diagnosis	1	1	0.75
Tanner at last visit	4 (1-5)	2 (1-5)	0.41
Age at start of puberty (years)	10.1 ± 2.9	9.2 ± 2.0	0.39
Height SDS at last visit	-0.97 ± 1.39	-0.72 ± 1.13	0.45
BMI SDS at last visit	1.08 ± 1.26	0.37 ± 1.02	0.02

Data is presented as n (%), mean ± SD, median (QIR)

Percentage of patients with one or more OPG-related endocrine comorbidities by NF1/non NF1



CONCLUSIONS

Patients with OPGs without NF1 had a higher rate of endocrine dysfunction presenting at a younger age compared to patients with NF1, which may be associated with tumor location and more aggressive treatments.