Childhood-onset Craniopharyngioma diagnosed as incidentaloma – Results of the prospective multicenter trial KRANIOPHARYNGEOM 2007

Svenja Boekhoff[1], Brigitte Bison[2], Maria Eveslage[3], Panjarat Sowithayasakul[4], Hermann L. Müller[1]

[1] Dep. of Pediatrics and Pediatric Hematology/Oncology, University Children's Hospital, Klinikum Oldenburg AöR, 26133 Oldenburg, Germany; [2] Dep. of Neuroradiology, University Hospital Würzburg, 97080 Würzburg, Germany; [3] Inst. of Biostatistics and Clinical Research, University of Münster, 48149 Münster, Germany; [4] Dep. of Pediatrics, Srinakharinwirot University, 26120 Bangkok, Thailand.

Background

Childhood-onset craniopharyngiomas (CP) are diagnosed due to clinical symptoms (symCP) or incidentally (incCP). We investigated clinical manifestations and outcome in incCPs and symCPs.

Patient cohorts

	symCP	incCP
Patients, *n*	214	4
Gender, male/female, *n* (%)	101 (47) / 113 (53)	3 (75) / 1 (25)
Age at diagnosis (years)	9.6 (1.3 – 17.9)	8.1 (3.7 – 15.2)
Duration of history (months)	5 (0 – 108)	/
Follow-up time (years)	3.4 (0.0 – 10.9)	5.9 (2.1 – 8.0)
BMI-SDS at diagnosis	0.48 (-3.82 – 10.02)	1.54 (-0.57 – 3.88)
Height-SDS at diagnosis	-1.01 (-4.90 – 3.64)	0.38 (-1.25 – 1.07)
BMI-SDS at last visit	2.60 (-2.89 – 13.22)	2.76 (2.08 – 8.00)
Height-SDS at last visit	-0.43 (-4.94 – 3.09)	-0.02 (-1.49 – 2.79)
Hydrocephalus at diagnosis, *n* (%)	83 (38.8)	0 (0)
Tumor location, *n* (%)		
Extrasellar location	44 (21)	2 (50)
Intra and extrasellar location	153 (72)	2 (50)
Intrasellar location	3 (1)	0 (0)
n.a.	14 (6)	0 (0)
Tumor size (cm²)	12.16 (0.004 – 79.54)	3.26 (0.56 – 5.13)
Hypothalamic involvement (HI)		
No HI	11 (5)	1 (25)
Anterior HI	55 (26)	1 (25)
Anterior and posterior HI	141 (66)	2 (50)
n.a.	7 (3)	0 (0)

Table 1: Characteristics in patients diagnosed and recruited with incidentaloma craniopharyngioma (incCP) and symptomatic craniopharyngioma (symCP) in the trial KRANIOPHARYNGEOM 2007 between 2007 and 2014.

Results

Reasons for imaging in incCP were cerebral palsy (case 1), head trauma (case 2), nasal obstruction (case 3), and tethered-cord syndrome (case 4), whereas headache (44%) visual impairment (25%), and growth retardation (17%) lead to imaging in 214 symCP. Tumor size at diagnosis was smaller in incCP when compared with symCP. Age, gender, BMI, height, hydrocephalus, tumor location, and hypothalamic involvement at diagnosis of incCP were within the range of these parameters in symCP. Complete resections were achieved more frequently in incCP when compared with symCP (20%). Surgical hypothalamic lesions were distributed similar in incCP and symCP. XRT was performed only in symCP (33%). No noticeable differences were observed concerning survival rates, endocrine deficits, BMI, height, functional capacity and QoL of the 4 incCP cases when compared with the symCP

MRIs of incidentaloma cases

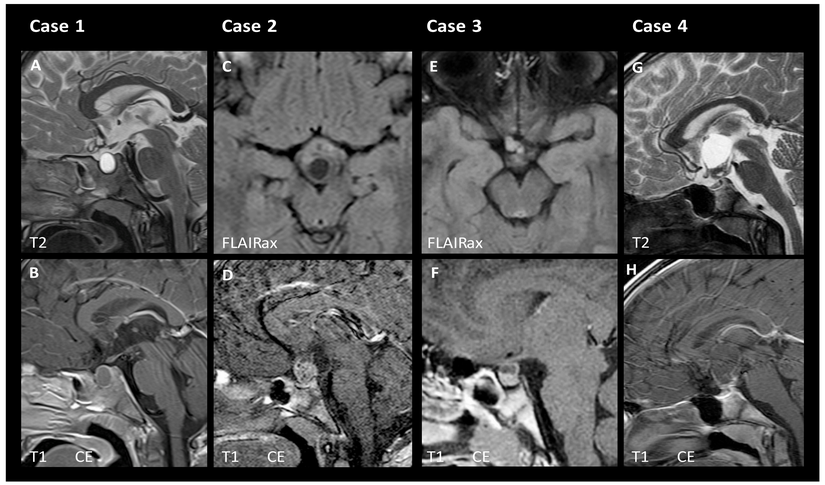

Figure 1: MRI at diagnosis in 4 patients (**Figure 1 A–H**) diagnosed and recruited with incidentaloma craniopharyngioma (incCP) in the trial KRANIOPHARYNGEOM 2007 between 2007 and 2014. **Figure 1A,B** (Case 1): Small, predominantly intrasellar and cystic lesion, displacing the sellar diaphragm upward, no contact to the optic system. **Figure 1C,D** (Case 2): Medium sized, partly solid, partly cystic lesion compressing the optic chiasm as well as the anterior and posterior hypothalamus, primarily located in the suprasellar region, edema of the tractus opticus. **Figure 1E,F** (Case 3): Small, partly solid, partly cystic lesion with an intra- and suprasellar location, displacing the optic chiasm slightly upward. **Figure 1 G,H** (Case 4): Medium sized, mostly suprasellar and cystic lesion with compression of the anterior and posterior hypothalamus. Additionally bleeding in a small Rathke's cleft cyst. In all cases, the solid parts and the cyst walls show some contrast enhancement and the cyst content some elevated signal on T1 due to protein-rich and colloid content.

Survival rates

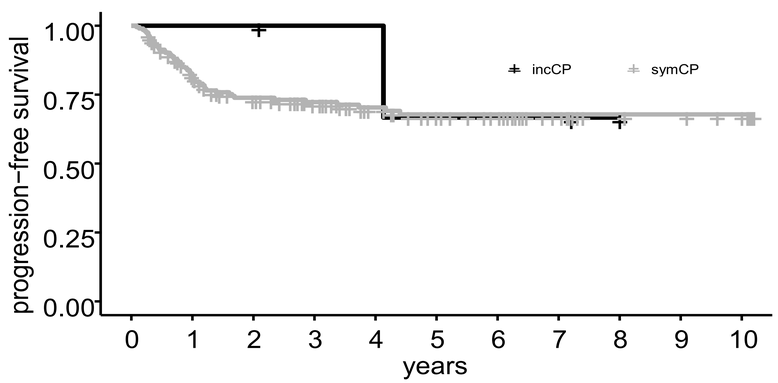

Figure 2: Kaplan-Meier progression-free survival rate (PFS) in patients diagnosed with incCP and symCP in the trial KRANIOPHARYNGEOM 2007 between 2007 and 2014.

Design and methods

IncCP were discovered in 4 (3m/1f) and symCP in 214 (101m/113f) CP recruited between 2007 and 2014 in KRANIOPHARYNGEOM 2007. Age, sex, height, body mass index (BMI), tumor size, degree of resection, pre and postsurgical hypothalamic involvement/lesions, pituitary function and outcome were compared between both subgroups.

Treatment characteristics

	symCP	incCP
Patients, n	214	4
Surgical approach, *n* (%)		
Transcranial approach	96 (45)	3 (75)
Transsphenoidal approach	32 (15)	1 (25)
Other approach	13 (6)	0 (0)
n.a.	73 (34)	0 (0)
Degree of resection, *n* (%)		
Complete resection	42 (20)	3 (75)
Incomplete resection	134 (63)	1 (25)
n.a.	38 (17)	0 (0)
Hypothalamic lesion (HL), *n* (%)		
No HL	62 (29)	1 (25)
Anterior HL	73 (34)	1 (25)
Anterior and posterior (HL)	70 (33)	2 (50)
n.a.	9 (4)	0 (0)
Irradiation, *n* (%)		
Irradiation	71 (33)	0 (0)

Table 2: Treatment characteristics in patients diagnosed and recruited with incidentaloma craniopharyngioma (incCP) and symptomatic craniopharyngioma (symCP) in the trial KRANIOPHARYNGEOM 2007 between 2007 and 2014. CP, craniopharyngioma; incCP, CP diagnosed as incidentaloma; symCP, symptomatic craniopharyngioma diagnosed due to clinical symptoms and complaints; HL, surgical hypothalmic lesion; n.a., data not available.

Conclusions

IncCP are rare (1.8%) and characterized by lack of endocrine deficiencies, resulting in normal height and BMI, no hydrocephalus, and smaller tumor size at diagnosis when compared with symCPs. Outcome of the observed incCP is similar with symCP.

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