SYMPTOMATIC RATHKE CLEFT CYST IN PEDIATRIC PATIENTS

clinical presentations, surgical treatment and postoperative outcomes - an analysis of 38 cases

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OBJECTIVES

Rathke cleft cysts are benign, epithelium-lined intrasellar and/or suprasellar cysts believed to originate from remnants of the Rathke pouch. Although its prevalence in adults is rather high, Rathke cleft cysts are rare in children. Often they are asymptomatic findings, however depending on their size and localization they can present with a wide spectrum of symptoms.

The aim of the study was to analyse the symptoms and surgical outcome of patients with the diagnosis of Ratke cleft cyst based on histopathological examination of postoperative material.

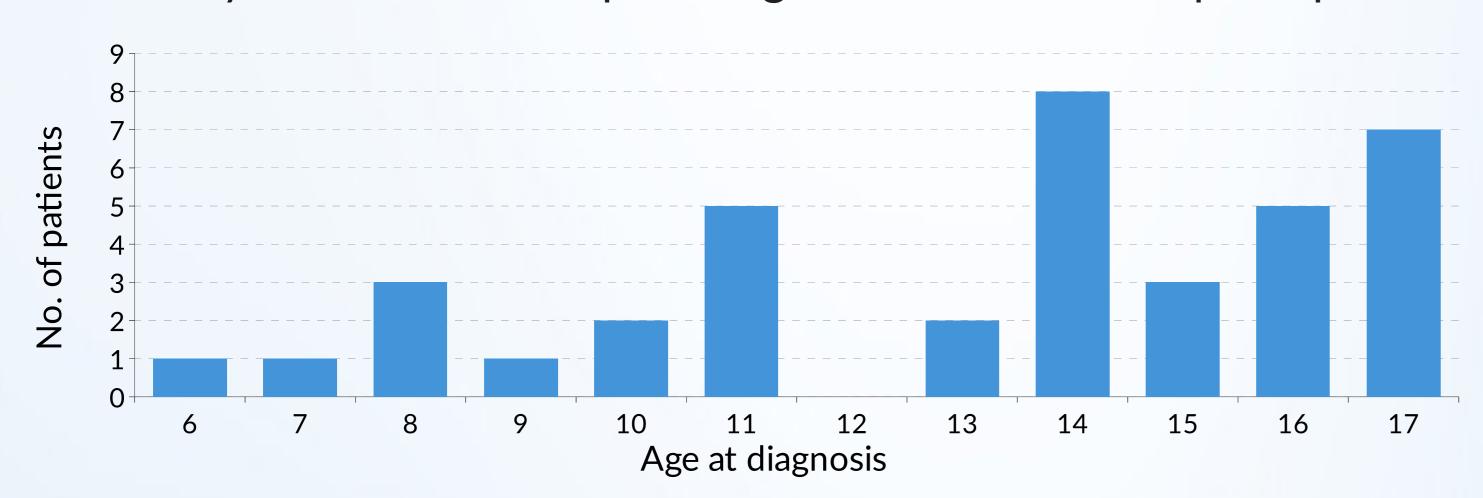


Fig.1. Age of patients at diagnosis

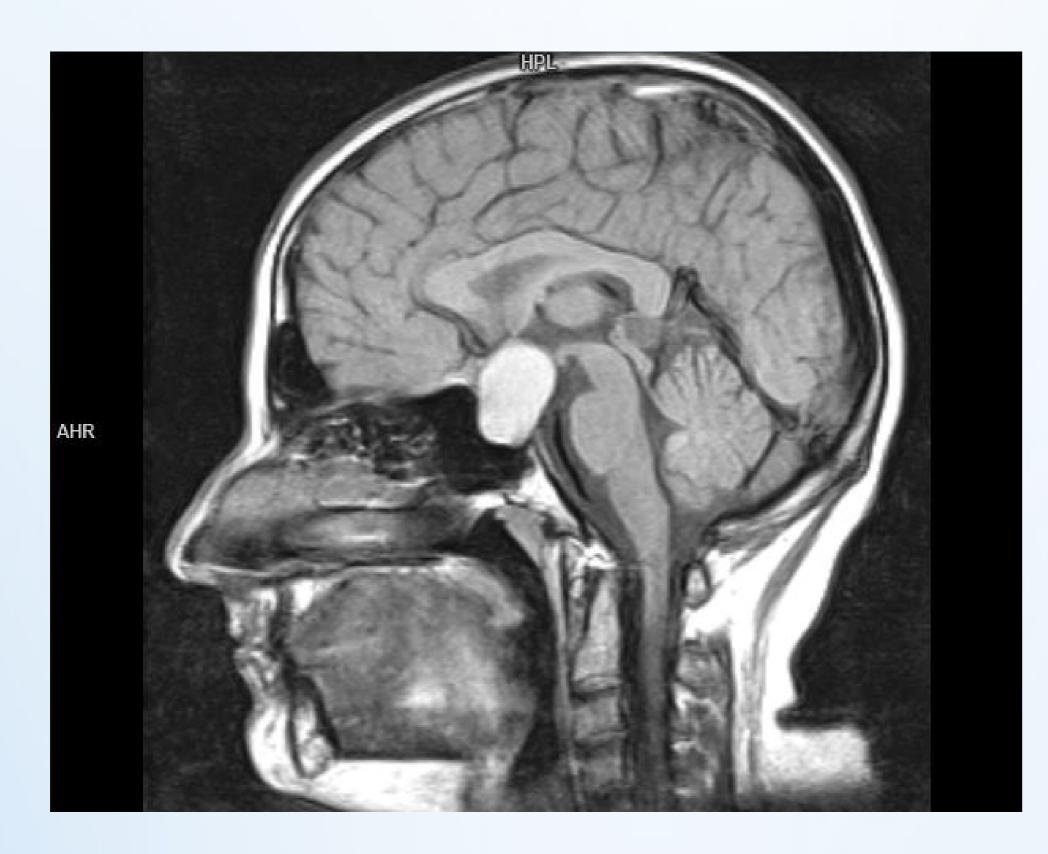


Fig.2. MRI image of patient with Rathke Cleft Cyst

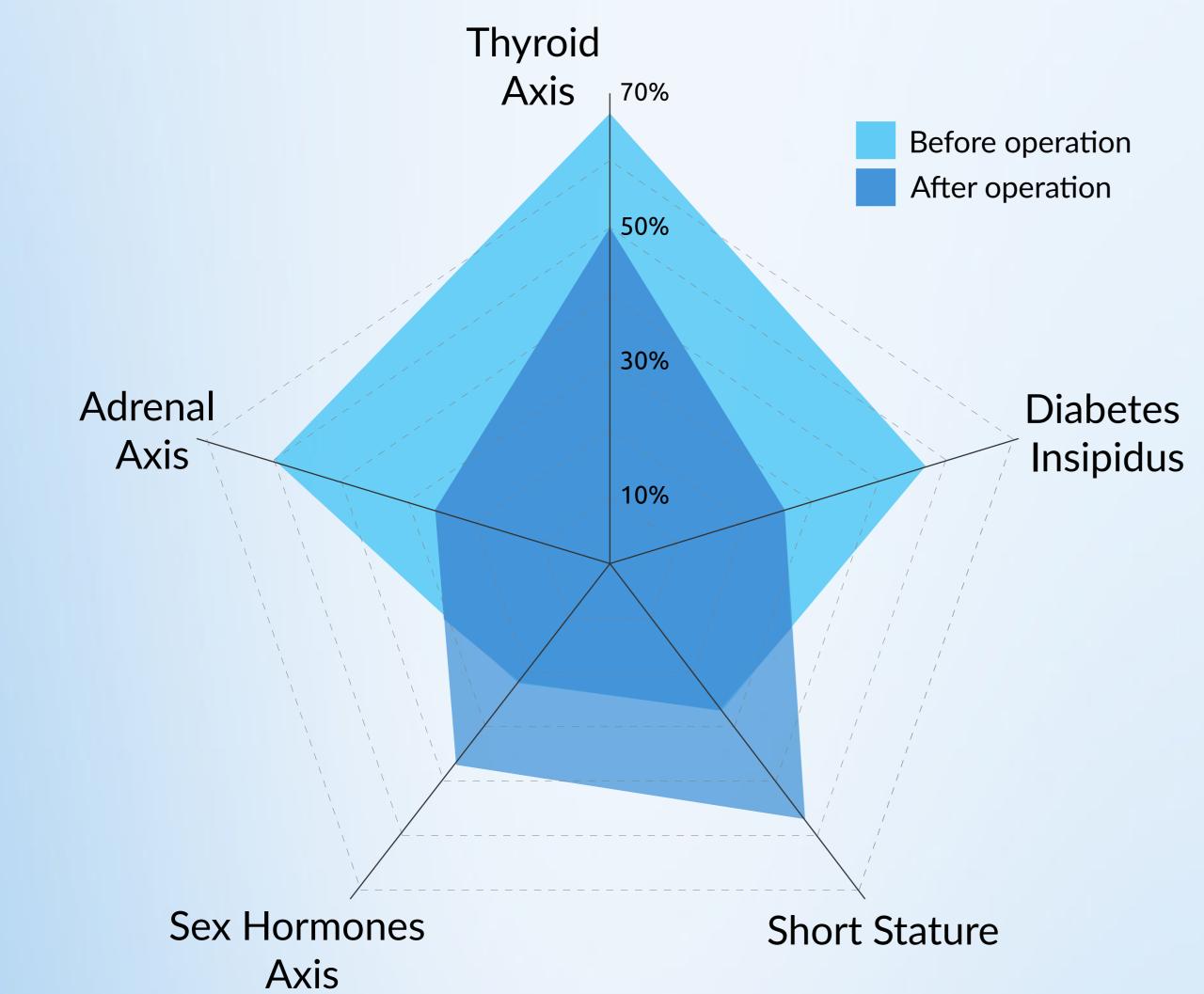


Fig.3. Deficits in hormonal axes before and after operation

METHODS

The study is a retrospective analysis of 38 cases of children who underwent a neurosurgical treatment due to Rathke cleft cyst in Children's Memorial Health Institute in Warsaw, Poland between 1994 and 2015.

RESULTS

Sex ratio was 1:0,9 (20 girls and 18 boys), two female patients were twins. At diagnosis patients were between 6 yrs. 11 mo. and 17 yrs. 10 mo. old with mean age of 13 yrs. 8 mo. [Fig.1.]

Average diameter of the cyst was 16,7 mm. In 6 cases calcifications in the lesions were observed either in brain imaging or intraoperatively.

Most common symptoms: headache (50%), hypothyroidism (50%), short stature and/or decreased growth velocity (47%), delayed puberty and menstrual abnormalities (37%), diabetes insipidus or polydipsia and polyuria (26%), adrenal dysfunction (26%), sleepiness and general weakness (13%), visual disturbances (11%). [Fig.3.]

29 patients underwent a transsphenoidal operation and in 9 a craniotomy was performed. All but one were successful (one patient died due to postoperative neurosurgical complications).

Follow-up of 30 out of 37 patients:

- average follow-up (time since operation until survey) is 8 yrs. 3 mo.
- average age of patients at the time of survey is 23 yrs. 9 mo. (20 adults and 10 below 18 yo).
- metabolic problems: 3 patients are obese (10%), 4 are overweight (13%), on the other hand 6 are underweight (20%). 2 obese patients are treated with metformin.
- education: 11 out of 13 patients (85%) over 25 yo have higher education.
 Among patients younger than 25 yo: 10 follow the regular school career (59%), 4 finished secondary school and entered working market (24%), 2 has individual education (11%) and 1 interrupted obligatory education (6%).
- pharmacological treatment: 6 patients don't need any at all (20%) and 2 of them are not under endocrinological care any more. Among the 24 that need pharmacological treatment 20 (67% of the entire group) need suplementation of thyroid axis, 15 (50%) of adrenal axis, 14 (47%) suffer from diabetes insipidus and obtain adequate treatment, 13 (22%) need suplementation of sex hormones and 8 (27%) are treated with growth hormone. [Fig.3.]
- 3 patients (10%) have subjective opthalmological problems (losses of visual field and in 1 case atrophy of the optic nerve).
- in case of 3 patients (10%) a reoperation due to Rathke cleft cyst was neccessary, which was conducted after 2, 5 and 11 years after first operation (on average 6 years).

CONCLUSIONS

Although Rathke clefts cysts are considered to be benign, non-neoplastic lesions they can present with serious symptoms that deteriorate significantly patients' quality of life. In spite of successful neurosurgical treatment in most of the analysed cases patients required a long-term pharmacological treatment due to endocrinological complications.

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