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Craniopharyngioma – case report and literature review

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Abstract

Craniopharyngiomas (Erdheim tumors) are rare tumors with low histological malignancy. Craniopharyngiomas occur at a rate of 0.5 to 2 cases per million people per year, accounting for 30 to 50% of all cases occurring in childhood and adolescence. Craniopharyngioma accounts for 1.2 to 4% of all intracranial tumors in children.

Clinical manifestations usually result from increased intracranial pressure, visual disturbances and endocrine disorders. Although long-term survival is high, patients' quality of life is poor, and patients' neuropsychological functions are often impaired. Choosing an appropriate treatment method, is a major challenge and requires the cooperation of physicians from different specialities.

Research priorities should be efforts to make information available on cases of Craniopharyngioma, as these tumors are extremely rare. The purpose of this article is to present a case of a boy with a tumor of the periventricular region based on the analysis of medical records and to analyze the current state of knowledge about craniopharyngioma.

Keywords: "*craniopharyngioma*", "*craniopharyngioma treatment*", "*craniopharyngioma diagnosis*".

1. Introduction

Craniopharyngiomas are rare neoplasms of low-grade malignancy (WHO I). This neoplasm is the most common intracranial tumor of non-glial origin (accounting for 5.6-14.1% of central nervous system tumors in children). There are two histopathological types of this tumor: adamantinomatous craniopharyngioma (ACP) and papillary craniopharyngioma (PCP). ACP is diagnosed with two peaks of incidence (5-15 years and 45-60 years), while PCP occurs only in adults, mainly in the fifth and sixth decades of life. The name "craniopharyngioma" was introduced by Cushing in 1932.

There are two hypotheses for the formation of craniopharyngioma:

1. it arises from the epithelial remnants of the craniopharyngeal duct or Rathke's pocket,
2. arises from metaplasia of residual squamous epithelial cells of the anterior lobe of the pituitary gland and funiculus [1, 2, 3, 4].

The most common location of the tumor is the saddle and peri-saddle region. Unfortunately, despite the high survival rate (85-95% 20-year survival rate), the quality of life for patients is poor [1, 2, 5, 6].

Typical symptoms of craniopharyngioma include:

1. increased intracranial pressure,
2. visual disturbances,
3. endocrine deficits.

The diagnosis of craniopharyngioma is often made several years after the first symptoms appear. Recent literature has documented that any clinical combination of headache, visual disturbances, decreased growth rate, and polydipsia or polyuria should raise the suspicion of pediatric craniopharyngioma and should be considered in the differential diagnostic process [1, 2, 7, 8, 9, 10].

For favorably localized craniopharyngioma, the preferred treatment of choice is to attempt complete resection with preservation of visual and hypothalamic function. For tumors with unfavorable localization, there are divergent opinions on the preferred method of treatment [11, 12, 13, 14].

2 Material and method

The purpose of this article is to present the case of a boy with a tumor of the periventricular region on the basis of an analysis of medical records and an analysis of the current state of knowledge about craniopharyngioma.

Using the PubMed platform, publications presenting diagnostic and therapeutic methods of craniopharyngioma were reviewed. The search included the keywords "craniopharyngioma", "craniopharyngioma treatment", "craniopharyngioma diagnosis".

3 Case description

A boy aged 7 years and 4 months was admitted to the hospital for a seizure, nausea and vomiting. A previously performed CT scan of the head without contrast showed abnormalities by which a suspicion of a neoplastic process was raised. On admission, the patient was conscious, oriented auto- and allopsychically. On neurological examination, nystagmus when looking to the right, positive Babinski sign on the right side, discrete central facial nerve palsy on the right side and weakness of the right upper limb were observed. A general pediatric examination showed no other pathology. Three days later, he underwent an MR scan of his head, which confirmed a proliferative process (Fig. 1.1a, 1.1b) The boy was qualified for an expedited neurosurgical operation. The procedure was performed via pterional craniotomy. On histopathological examination, the diagnosis was made - craniopharyngioma (Latin: craniopharyngioma, ademantinomus type WHO GI). After the surgery, the patient's symptoms of palsy syndrome persisted, in addition, hypopituitarism and features of

oculomotor nerve palsy on the left side were found. One month after the operation, the boy was admitted to the Neurology Department for postoperative consultation. A CT scan of the head with contrast was performed, which showed no features of CNS hemorrhage, as well as a follow-up MR study (Fig. 1.2). Drug treatment was prescribed, and a visit to the Rehabilitation Department and follow-up examinations were scheduled. One month later, a follow-up CNS CT scan was performed, which showed significant regression of the left-sided cerebral hydrops and progression of ventricular system dilatation. Four months after surgery, a follow-up MR study was performed (Fig. 1.3). Two weeks after the last follow-up visit, the patient presented to the ED with a severe headache. Physical examination revealed right-sided spastic paresis. A head CT scan was performed, which showed progression of ventricular system dilatation.

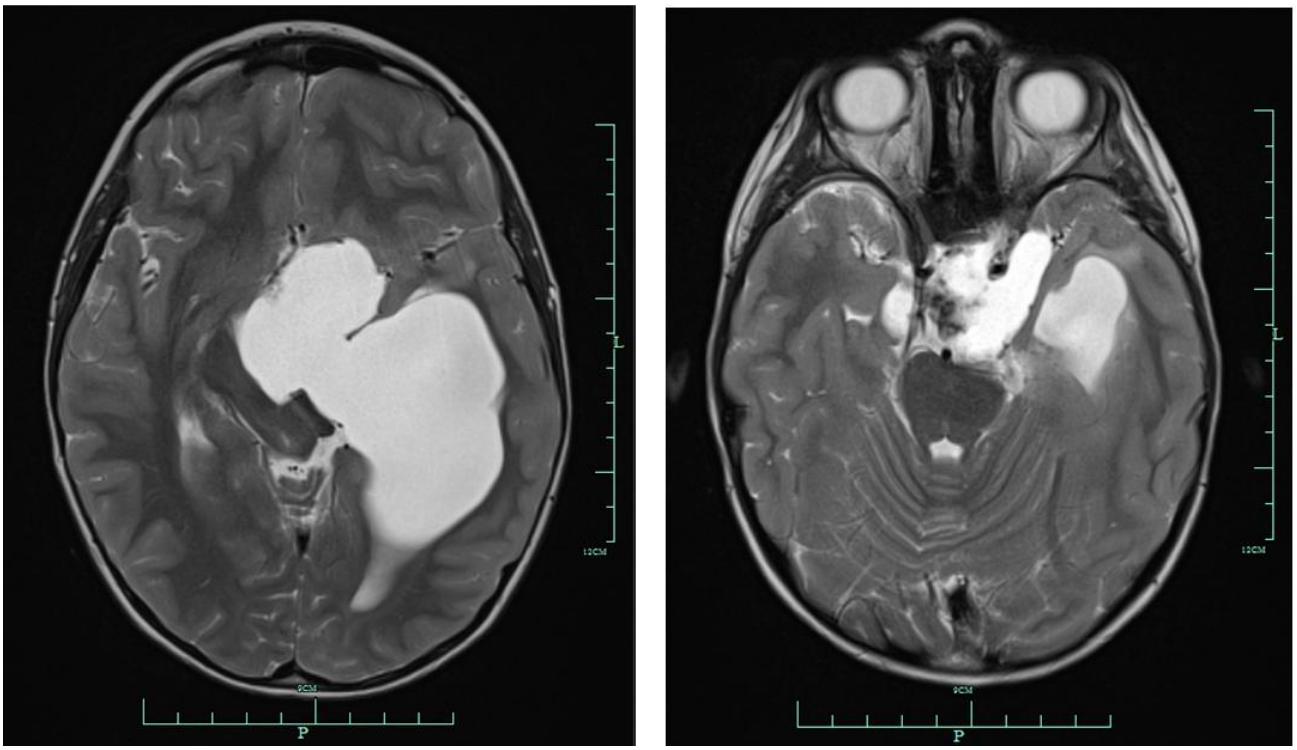

Fig. 1.1.a, 1.1.b MR image

Suprasellar medially and on the left side, an extensive cystic lesion of 81 x 93 x 41mm (RL x AP x CC) in the largest cross-section is seen displacing the midline structures by about 15mm to the right side, compressing the left optic nerve, the optic chiasm and the ventricular system.

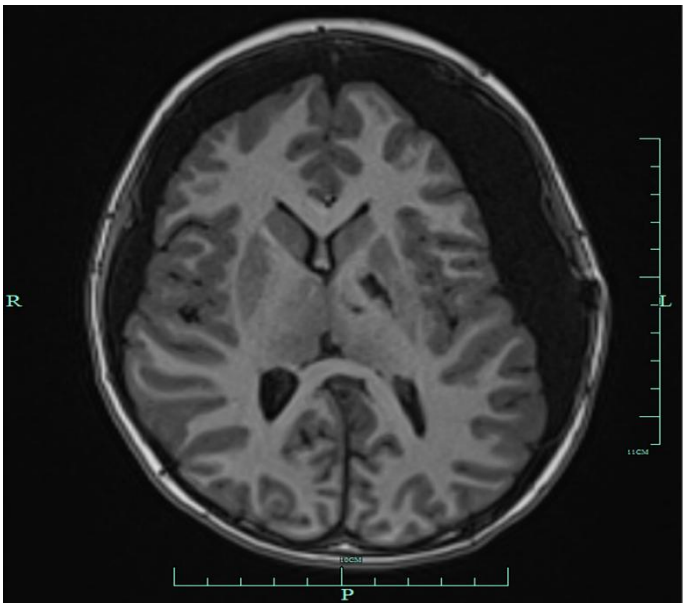

Figure 1.2: MR image.

Condition after craniopharyngioma surgery. Bilateral parenchymal hydrocephalus and midline displacement of about 4mm are visible.

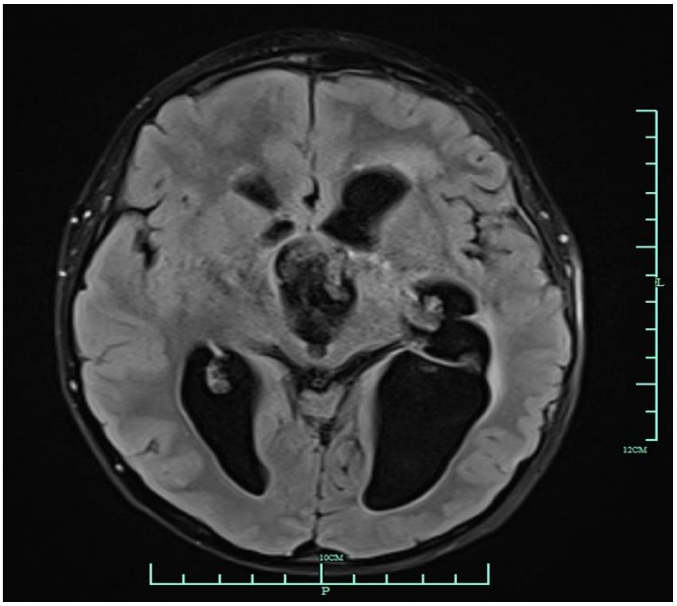

Figure 1.3: MR image.

Complete regression of the parocerebral hydrops. Quite significant asymmetric dilatation of the lateral ventricular system and the third ventricle can be seen.

4 Discussion

4.1. Pathogenesis of craniopharyngioma.

The pathogenesis of craniopharyngioma is not precisely known. Currently, two theories have been posited to explain the pathogenesis of the tumor - embryonic and metaplastic.

The embryonic theory is that craniopharyngiomas arise from the neoplastic transformation of embryonic nests of squamous cells of the involutional craniopharyngial duct. During the process of proliferation and turnover of Rathke's pouch cells, the remnants of the cells of the craniopharyngeal duct spread into the midline and suprasellar region.

The metaplastic theory tells us that craniopharyngiomas arise from metaplasia of residual squamous epithelial cells of the anterior lobe of the pituitary gland and funiculus, resulting in nests of squamous cells.

4.2. Symptoms of craniopharyngioma.

The symptoms of craniopharyngioma are highly variable. The location of the tumour has a direct impact on the symptoms. At the time of diagnosis, the patient's clinical picture is often dominated by non-specific symptoms of increased intracranial pressure. Other common symptoms are visual disturbances (62-84%) and endocrine deficits (52-87%). Endocrine disorders are due to a disruption of the hypothalamic-pituitary axis. An early sign of craniopharyngioma is a pathologically reduced growth rate. Significant weight gain, indicative of hypothalamic obesity - usually occurs later, shortly before diagnosis. At the time of diagnosis, 40 to 87% of patients are found to have at least one hormone deficiency, such as hypopituitarism. Other endocrine manifestations, such as neurohormonal uremia, are present before surgery in 17 to 27% of patients [1, 15, 16, 17].

Hoffmann et al. observed that the median duration of pre-diagnosis history was approximately six months, ranging from 0.1 to 108 months, and was positively correlated with the patient's age at diagnosis. Tumour size, degree of surgical resection,

hypothalamic involvement and body mass index (BMI) at diagnosis were not associated with disease duration. Endocrine deficiencies at diagnosis were associated with long disease duration Neurological deficits and visual impairment were associated with larger tumour size at diagnosis and reduced 10-year overall survival [18].

The described case presented to hospital due to non-specific symptoms - seizure, nausea and vomiting. These may be due to an increase in intracranial pressure. Head imaging studies are important in the diagnostic process. An MRI scan of the head confirmed a proliferative process, which prompted the doctors to qualify the patient for fast-track neurosurgery.

4.3. Diagnosis of craniopharyngioma.

Imaging studies have shown that a craniopharyngioma is usually a cystic tumour of the intrasacral and suprasacral region. The most common location of the tumour is the supra-sacral region, with an intra-sacral portion. In contrast, 20% are exclusively suprascapular and 5% exclusively intrascapular. Contrast-enhanced MRI is the standard imaging study for the detection of craniopharyngioma. It is also worth noting the importance of CT scans performed to detect or exclude calcifications in craniopharyngioma tissue, which are found in 90% of these tumours [19, 20].

Differential diagnoses include hypothalamic and optic nerve glioma, Langerhans cell histiocytosis, Rathke's cleft cyst, yellow granuloma, intracranial germinoma, epidermoid tumour, arachnoid cyst thrombosis, third ventricle colloid cyst, pituitary adenoma, aneurysm and inflammatory lesions [20, 21].

It is worth mentioning that after tumour resection, a postoperative head CT scan is recommended to definitively confirm complete resection. Imaging to detect recurrence or progression during follow-up should be limited to MRI to increase diagnostic sensitivity and reduce radiation burden [20].

4.4. Treatment of craniopharyngioma.

Treatment may include surgery alone, radiation alone or a combination of the two. Surgery appears to be appropriate for tumours that can be completely resected, preferably without damage to the optic nerves or hypothalamus. Avoiding irreversible damage to the hypothalamus is a key goal in treatment. Studies show that success in choosing the

correct surgical method, as well as any subsequent surgical complications during resection, are linked to the experience of the surgeon [22, 23, 24]. Tumours originating from the pituitary peduncle and tumours extending into the funiculus can be reached by the classical pterional or infra-pituitary route [25, 26, 27].

Surgery may also be performed to alleviate symptoms or improve the patient's ability to tolerate irradiation [22]. Limited surgery can take various forms. In all cases of incomplete tumour removal during transcranial surgery, catheters connected to, for example, a Rickham reservoir can be inserted into the remaining cysts for subsequent aspiration of fluid from the cyst or infusion of obli terative substances. It should be noted that aspiration or fenestration of the cyst may have a temporary benefit but reaccumulation of fluid during irradiation is highly likely [22, 25, 26].

Irradiation refers to external beam radiotherapy using photons or protons. In most cases, radiotherapy is given at the time of progression, following previous surgery. Progression may result from unexpected recurrence of the disease after complete resection of the tumour. Irradiation is also used in the situation of incomplete resection [22, 28].

5 Conclusions

Craniopharyngioma is a tumour with a very diverse clinical picture. The diagnostic and therapeutic process is very challenging and requires the cooperation of doctors with different specialisations. Despite the absence of histological malignancy, it causes many dangerous symptoms that significantly reduce quality of life due to the sequelae caused by the tumour's anatomical proximity to the optic nerve/scleral and hypothalamic-pituitary axis. Due to its intracranial location, its radical treatment can be difficult and fraught with the risk of complications. The controversy surrounding the best treatment strategy for craniopharyngioma has not been resolved in the published literature to date.

Author's contribution

Conceptualization, Michał Leśniewski; methodology, Michał Leśniewski; software, Michał Leśniewski; check, Michał Leśniewski; formal analysis, Michał Leśniewski; investigation, Michał Leśniewski; data curation, Michał Leśniewski; writing – rough preparation, Michał Leśniewski; writing – review and editing, Michał Leśniewski; supervision, Michał Leśniewski; project administration, Michał Leśniewski; receiving funding, Michał Leśniewski.

All authors have read and agreed with the published version of the manuscript.

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Not applicable

Acknowledgments

Not applicable

Conflict of Interest Statement

The authors report no conflict of interest.

Supplementary materials

Fig. 1.1.a, 1.1.b MR image

Figure 1.2: MR image.

Figure 1.3: MR image.

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