Submitted: 22.11.2024
Accepted: 04.01.2025
Early publication date: 04.03.2025

Endokrynologia Polska
DOI: 10.5603/ep.103701
ISSN 0423–104X, e-ISSN 2299–8306

Aggressive craniopharyngioma with problematic course

Miłosz Chwiałkowski, Grzegorz Zieliński

Neurosurgery Department, Military Institute of Medicine — National Research Institute, Warsaw Poland

Keywords: *craniopharyngioma; pituitary; hypothalamus*

Craniopharyngiomas are rare slow growing, benign tumours histologically divided into 2 subtypes : adamantinomatous and papillary. Papillary craniopharyngioma is more common in adults with *BRAF V600E* mutation. The 2 main treatment methods are neurosurgical resection and radiotherapy [1, 2].

A 39-year-old female presented with aggressive craniopharyngioma in the parasellar region with significant enlargement in control study after 5 months, revealed during the diagnostic process for headaches. A magnetic resonance imaging (MRI) scan showed a mixed tumour — cystic and solid component, 2 cm in diameter, with hypothalamus, pituitary stalk, third ventricle, and basilar artery involvement. In pre-operation assessment there were no diabetes insipidus or anterior pituitary lobe hypofunction detected, but with slight enlargement of the ventricles.

The patient was operated by the right side subtemporal approach. After subtotal resection of the tumour we observed DI and hypofunction of the anterior pituitary lobe and infarction of the left hypothalamus in a control CT scan. The patient was treated with desmopressin, levothyroxine, and hydrocortisone and was kept under constant physiotherapeutic care. Due to postoperative progression of hydrocephalus in control tests and deterioration of neurological status, a ventriculoperitoneal valve was implemented. Additionally, the patient has paresis of the oculomotor nerve, gait imbalance, and psychomotor sluggishness. In a control MRI scan after 2.5 months we observed complete growth of the tumour. In histological examination positive papillary craniopharyngioma BRAF V600 was described without anaplasia or any other malignant features. Although as the clinical condition was severe, further treatment was aborted, including radiotherapy. The patient was discharged home 2.5 months postoperatively.

Craniopharyngioma is a challenging, locally aggressive parasellar tumour with a rate of less than 5% among all intracranial tumours. It is one of the most complicated tumours in neurosurgical practice due to its localisation near vital neurovascular structures, e.g. the hypothalamic-pituitary axis. It is a significant challenge to preserve all the small perforating arteries during surgery because even small manipulation can lead to hypothalamus infarction, such as in our case. The frequency of recurrence varies, but in general this tumour grows slowly. In our case we observed very rapid growth in 2 months; however, resection was not complete. Nevertheless, in subtotal resection, depending on the study, relapse is observed between 1 and 11 years. Despite the operation, one of the most difficult problems is postoperative care. Frequently, especially in pituitary stalk involvement, after the surgical procedure multihormonal hypoactivity and electrolyte disorders are observed, as in our case. In order to rebalance this, a close assessment of liquid balance, and sodium and potassium levels is required. Determination of the appropriate level of desmopressin dose demands time and patience. Quick substitution of hydrocortisone is mandatory. Hydrocephalus is another important problem generally demanding resolution before tumour excision. We observed no significant bleeding during the operation, and it was confirmed in control tests such computed tomography (CT) or MRI scan, which could be responsible for developing hydrocephalus postoperatively. The reason for this complication is unclear. Our experience proves that it is not a principle to resolve it earlier, but it is necessary to monitor it and engage in appropriate decisions depending on the circumstances [3–5]. Histological examination confirmed papillary craniopharyngioma with BRAF

Miłosz Chwiałkowski, Neurosurgery Department, Military Institute of Medicine — National Research Institute, Szaserów 128 St., Warsaw, Poland; e-mail: mchwialkowski@wim.mil.pl

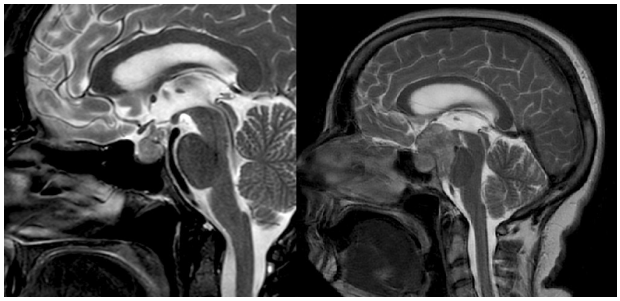

Figure 1. *In order: first study during diagnostic process, second picture — 5 months after first magnetic resonance imaging (MRI)*

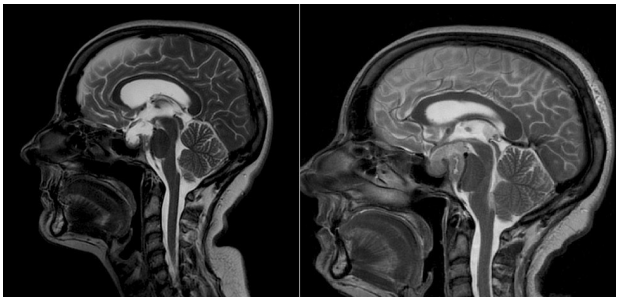

Figure 2. *In order: control study 7 days after operation and 65 days after operation*

V600 expression where pharmacological treatment with BRAF/MEK inhibitors could be implemented, but the availability of this form of therapy is currently limited [3]. Such rapid progression without histological features of tumour anaplasia, classified according to the World Health Organisation (WHO) as GI, is worrying. The current qualification allows for the use of the above-mentioned inhibitors in treatment in the future, which has not been used so far.

Attention to patients with craniopharyngioma should remain in highly specialised multidisciplinary centres due to high morbidity rates, such as from hypopituitarism, cardiovascular risk, hypothalamic damage, visual and neurological symptoms, and neurocognitive function.

Conflict of interests

The authors have no conflicts of interest to disclose.

Statements of ethics

The research was conducted ethically in accordance with the World Medical Association Declaration of Helsinki. Data were collected retrospectively.

Funding

No funding was available.

References

1. Gao S, Shi X, Wang Y, et al. Malignant transformation of craniopharyngioma: case report and review of the literature. J Neurooncol. 2011; 103(3): 719–725, doi: 10.1007/s11060-010-0407-2, indexed in Pubmed: 20872276.
2. Liubinas S, Munshey A, Kaye A. Management of recurrent craniopharyngioma. J Clin Neurosci. 2011; 18(4): 451–457, doi: 10.1016/j.jocn.2010.10.004, indexed in Pubmed: 21316970.
3. Agosti E, Zeppieri M, Antonietti S, et al. Advancing Craniopharyngioma Management: A Systematic Review of Current Targeted Therapies and Future Perspectives. Int J Mol Sci. 2024; 25(2), doi: 10.3390/ijms25020723, indexed in Pubmed: 38255797.
4. Erfurth EM, Holmer H, Fjalldal SB. Mortality and morbidity in adult craniopharyngioma. Pituitary. 2013; 16(1): 46–55, doi: 10.1007/s11102-012-0428-2, indexed in Pubmed: 22961634.
5. Honegger J, Tatagiba M. Craniopharyngioma surgery. Pituitary. 2008; 11(4): 361–373, doi: 10.1007/s11102-008-0137-z, indexed in Pubmed: 18636330.