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Case Report

Surgical treatment of a rare rosette-forming glioneuronal tumor in the pineal region

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ABSTRACT

Background: Rosette-forming glioneuronal tumors (RGNTs) in the pineal region are rare. RGNTs have been described in the fourth ventricle, but rarely reported in other brain regions. Here, we report the radiological findings, surgical treatment, and short-term outcome of an RGNT found in the pineal region.

Case Description: We present a case of a 22-year-old medical student with a 4-month history of headaches and diplopia. A previous magnetic resonance imaging scan revealed a mass in the pineal region, with heterogeneous contrast enhancement and hydrocephalus. Three months prior, an endoscopic biopsy and third ventriculocisternostomy were performed elsewhere; the diagnosis was neurocytoma Grade I, and radiotherapy was planned. The patient presented at our hospital for a second opinion, and we suggested surgical treatment. A near-total resection was performed in sitting position using a supracerebellar infratentorial microsurgical approach. The tumor was very soft and not well vascularized. Diplopia was initially worsened after the tumor was removed and relieved completely after 2 weeks. An 8-week follow-up examination revealed that the patient was free of symptoms. Histological analysis confirmed it was an RGNT.

Conclusion: Maximal safe resection in pineal region RGNTs is a feasible and recommended treatment option.

Keywords: Operative nuances, Pineal region, Rosette-forming glioneuronal tumor

INTRODUCTION

Rosette-forming glioneuronal tumors (RGNTs) are an unusual variant of mixed neuronal-glia tumors. RGNTs are typically found in the fourth ventricle, with limited extension to the surrounding structures (cerebellar vermis, midbrain, and cerebral aqueduct). They were initially classified as novel “rosette-forming glioneuronal tumors of the fourth ventricle” according to the 2007 WHO classification of central nervous tumors.^[9] However, an increasing number of case reports have described these tumors in various locations beyond the fourth ventricle, including cerebellar hemispheres, vermis, chiasm, pineal region, third ventricle, hypothalamus, and spinal cord.^[3,5,7,11,13] Hence, in 2016, the revised version of the WHO classification described these tumors as “rosette-forming glioneuronal tumors” (without reference to the fourth ventricle), histologically Grade I.^[10] Around 150 cases of RGNTs have been described in the literature. However, less than 10 of these cases (5.3%) have been described in the pineal region.^[14] Pineal region RGNT may cause similar symptoms as the other pineal region tumors. However, microcalcifications and satellite lesion are

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commonly found in pineal region RGNTs. Characteristic features of RGNTs have recently been review by Anyanwu *et al.*^[2] Here, we present another case of an RGNT in the pineal region, which was surgically treated at our hospital.

CASE HISTORY

A 22-year-old medical student presented with a 4-month history of headache and diplopia. A previous magnetic resonance imaging (MRI) scan revealed a contrast-enhancing lesion in the pineal region. An endoscopic biopsy and third ventriculocisternostomy were performed elsewhere; the diagnosis was neurocytoma Grade I. Radiotherapy was planned elsewhere. The patient presented to our hospital for a second opinion. After oncological consultation, we suggested surgical resection of the tumor. On examination, the patient was alert and oriented. Visual field was intact, and pupil size was normal and reactive to light. The patient's hearing and facial nerve function were normal. A new MRI scan revealed a contrast-enhancing space-occupying lesion of approximately 2 cm in size in the pineal region [Figure 1]. No other pathology and no drop metastasis in the supra or infratentorial region were observed in preoperative workup.

Surgery of pineal region RGNT

Operative findings

Surgery was performed with the patient in sitting position. A supracerebellar infratentorial approach was

used, with an OPMI PENTERO 900 microscope (Carl Zeiss, Germany). The dura was opened in a Y-shaped fashion, preserving the main veins in the tentorial region. The tumor was easily recognized; it was soft, cystic, and pale yellow in color [Figure 2]. The tumor had a well-defined plane and was removed easily using standard microsurgical tools and techniques. The dura was closed in a watertight fashion.

Postoperative outcome

Diplopia was initially worse but completely relieved 2 weeks after the surgery, without any new focal neurological deficit. No other complications occurred. The patient was discharged from the hospital 4 days after the surgery. An 8-week follow-up examination revealed that the patient was without symptoms. Histological analysis showed an RGNT. Postoperative MRI scans showed a small remnant in posterior part of thalamus and aqueduct [Figure 3]. After discussion in interdisciplinary tumor board, the patient will be followed up with MRI scans.

DISCUSSION

An RGNT in the pineal region is a rare affliction. It mainly affects young adults – predominantly females – with a peak incidence among people in their thirties.^[8,14] However, our case report presents a 22-year-old male patient. The presentation of symptoms depends on where the tumor is located in the ventricular system or brain parenchyma. The

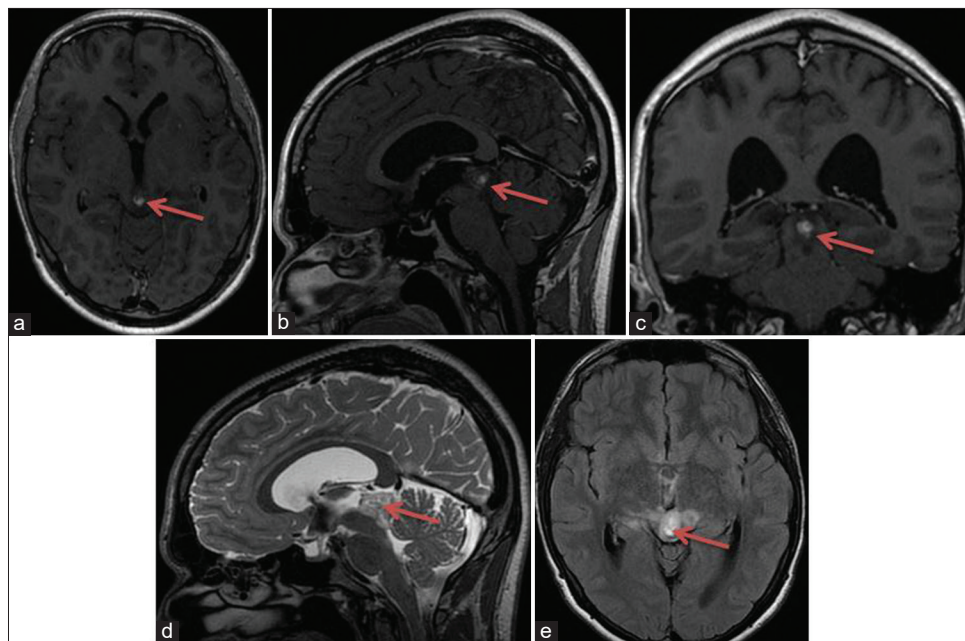


Figure 1: MRI scans (pre-operative) showing tumor location and radiological anatomy of a space-occupying lesion with partially and heterogeneous contrast enhancement in the pineal region (a,b,c). FLAIR and TW2 hyperintensity in and around the pineal region (d, e).

most commonly presented symptoms in RGNT cases are ataxia, vertigo, cranial nerve palsies, complaints of impaired vision, headache, and vomiting. Our patient presented with headache and diplopia. Radiologically, RGNTs can be cystic (35%), cystic solid (18%), or solid (47%). The solid tumor regions display hypointensity on T1W1 and hyperintensity on T2W2 and a variable but mostly heterogeneous pattern of contrast enhancement. In MR spectroscopy, the low-grade nature of these tumors is indicated by

slightly elevated choline, reduced N-acetyl aspartate, and the absence of lactate and lipids.^[4] Although we did not perform MR spectroscopy, the MRI images showed similar T1W1, T2W2, fluid-attenuated inversion recovery, and contrast enhancement as described in the literature. Multimodal neuroimaging may help to differentiate RGNTs preoperatively. Hydrocephalic symptoms resolved after the surgical resection in most of the cases in literature similar to our case. Histological analysis revealed a biphasic neurocytic and glial pattern with an absence of atypia, mitotic activity, necrosis, and low proliferation index, characteristic of RGNTs.

Due to limited experience with therapeutic treatments such as stereotactic radiosurgery or chemotherapy, surgical resection aimed at achieving gross total resection remains the preferred treatment of RGNTs. RGNTs are Grade I benign tumors, but aggressive progression and ventricular dissemination have been previously described.^[1,6,12] The risk of recurrence may be increased if resection is inadequate, and/or if purely solid tumors are found in patients of pediatric age. Due to shorter follow-up periods described in the literature and due to few cases of pineal RGNTs, it is difficult to draw a conclusion on the prognosis of RGNTs in pineal region. Hence, a long-term closely monitored follow-up is important. Genetic analysis of mutations, including IDH1 and IDH2, as well as chromosomal codeletion analysis may be useful in further characterization of these tumors.

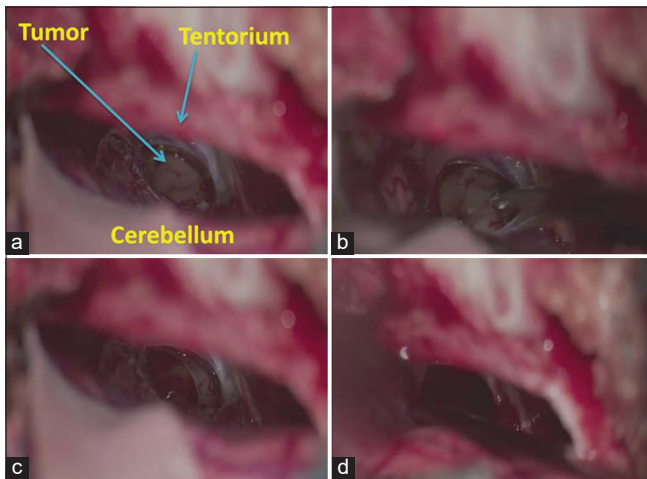


Figure 2: Intraoperative images showing tumor removal and intraoperative surgical anatomy (a-d).

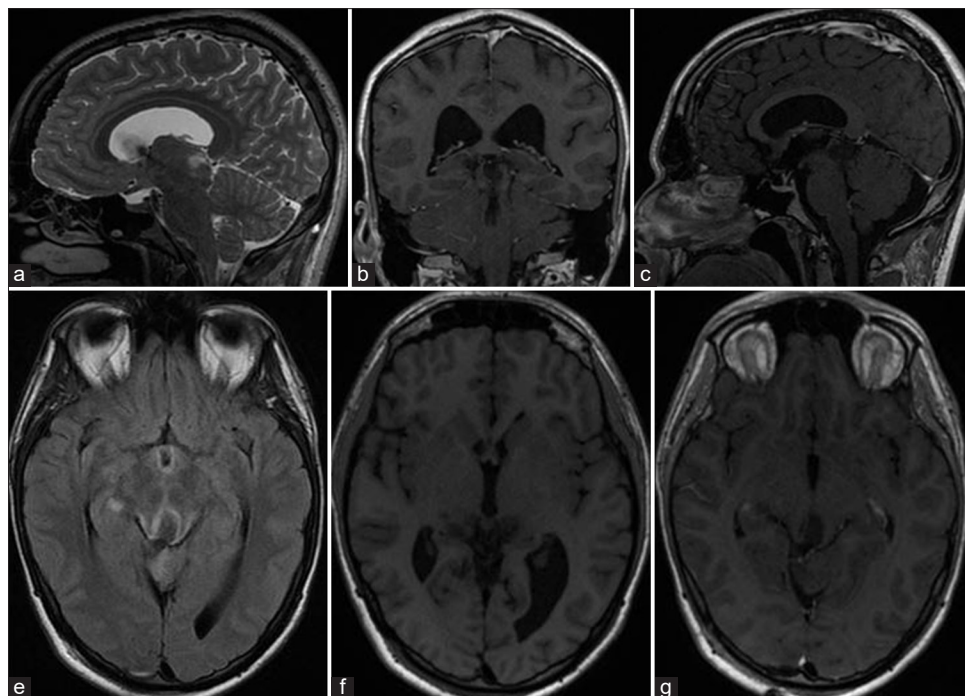


Figure 3: MRI scans (post operative) showing near total tumor removal (a, b, c, e and f). FLAIR (e) and TW2 (a) show slight signal enhancement (small tumor remnant) in posterior part of thalamus and aqueduct.

CONCLUSION

Maximal safe resection of RGNTs in the pineal region is a feasible and recommended treatment option.

Ethical approval

All procedures performed in this study involving human participants were in accordance with the ethical standards of the research committee of University of Helsinki and with the 1964 Declaration of Helsinki and its amendments or comparable ethical standards.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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