

Adult Supratentorial Extraventricular Anaplastic Ependymoma: Therapeutic Approach and Clinical Review

Abstract

We report a 69-year-old patient with left paresthesia and hemiparesis. Magnetic resonance imaging revealed a right frontoparietal cystic tumor. A subtotal surgical resection was performed, and an Ommaya reservoir was left in place. The pathological diagnosis was supratentorial extraventricular anaplastic ependymoma. Radiation therapy was administered, and Ommaya reservoir drainages were performed. Four months after, her clinical status deteriorated after a reservoir drainage and image revealed an acute hemorrhage. An additional resection was carried out, and chemotherapy was undergone. One month later the tumor relapsed and the patient died 18 months after initial diagnosis. Some poor prognostic factors have been suggested in the literature: Young age, incomplete tumor resection – eloquent area location, histological anaplasia, supratentorial, and extraventricular locations. Ommaya reservoirs may be used in cystic lesions as a temporary measure only. Surgery is the mainstay of therapy with adjuvant radiotherapy and/or chemotherapy.

Keywords: Chemotherapy, Ommaya reservoir, radiotherapy, supratentorial extraventricular anaplastic ependymomas, surgery, treatment

Introduction

Ependymomas are primary tumors of the central nervous system arising from the ependymal cells lining the ventricular system and central canal of the spinal cord. They account for about 2% of all intracranial tumors; most commonly affect the pediatric population, and in the adult represent 3–5% of the glial tumors.^[1-4] Although the majority of ependymomas develops infratentorially, they may also arise at the supratentorial level and rarely with no connection with the ventricular system.^[2,5,6] Three hypothesis have been proposed:^[7] Tumors develop from intraparenchymal or subarachnoid ependymal cysts originating from germinal matrix migration disorders; or they generate from neuroectodermal neoplasms that differentiate into the ependymal lineage; or, finally, they result from neoplastic growth of an ectopic ependymal cell, as a consequence of a migrational error.

Although the standard approach for anaplastic ependymomas is maximal surgical resection followed by postoperative focal irradiation in children,^[8] it is not clear whether pediatric population management

should be used in the adult supratentorial extraventricular counterpart.^[4]

Case Report

A 69-year-old woman presented with a left upper limb paresthesia and weakness to the emergency room. Neurological examination disclosed grade 4 paresis of the left upper limb and left hemihypesthesia. The magnetic resonance imaging (MRI) showed a right frontoparietal lesion with cystic and solid components [Figure 1]. A right parietal craniotomy was performed with resection of the mural nodule. Considering the eloquent area and the thin cyst wall with no anatomical cleavage plan, no gross total resection was possible. An Ommaya reservoir was inserted. Postoperative MRI showed mild enhancement of the cystic wall and decreased mass effect [Figure 2]. The pathological diagnosis was supratentorial extraventricular anaplastic ependymoma (SEAE) [Figure 3].

External conformational radiation therapy was administered in 30 daily fractions up to a total of 60 Gy.

The patient experienced some symptomatic deteriorations related to cystic fluid

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content controlled with Ommaya reservoir percutaneous drainage. After one episode, the patient did not improve, and an imagiological evaluation was performed sustaining signs of acute hemorrhage within the lesion cavity after drainage [Figure 4]. Clinical evaluation showed symptomatic worsening with a left hemiparesis.

The patient was reoperated but refused an aggressive surgical resection due to the high risk of hemiplegia. The cystic content was drained, and subtotal resection of the mural nodule and cystic walls were accomplished, preserving motor function. Postoperative MRI showed a linear contrast-enhanced zone and a 12.9 mm nodule remaining [Figure 5]. Histological examination still discloses some areas of typical ependymoma although most of the parenchyma tumor now presented features of an undifferentiated, high-grade glioma.

The patient partially improved being autonomous at hospital discharged. She was started on chemotherapy with temozolomide which she did not tolerate due to pancytopenia.

Her general condition deteriorated, she became partially dependent in daily living activities and died 18 months after the initial diagnosis, 9 months after reoperation.

Discussion

To the best of our knowledge, only 17 cases of adult intra-axial SEAE have been previously reported in PubMed [Table 1].^[4,5,9-16] The age of diagnostic was 18–70 years (mean = 39.4 ± 18.5 years). Our patient was 69 years old, an unusual age for this diagnosis as she is in the upper tail of age distribution. No meaningful differences according to gender were noted. Regarding the topographic tumor distribution, the frontal and parietal locations were the most frequent, being responsible for 67% (12/18) of the cases, and correlated with the presenting symptoms. The imagiological appearance of a subcortical cyst with a mural nodule is common in low-grade gliomas.^[17] However, supratentorial anaplastic ependymomas most frequently appear as a heterogeneous enhancing solid or cystic/solid lesion^[18] which is supported by this report. Despite some cases present hemorrhage and calcifications, none was present in this case. Surgery resection was performed in all the cases with data concerning treatment; 75% (12/16) of patients underwent radiation therapy; and 31% (5/16) adjuvant chemotherapy.

A reoperation was also performed due to a hemorrhagic event. In only 2 cases,^[13,16] reoperation is reported in the literature at the time of first recurrence. During the follow-up, 44% (8/18) of the tumors did not recur, 33% (6/18) had at least one episode of recurrence but for 22% (4/18) cases no data concerning recurrence exists. The minimum survival was 14 months with 44% (8/18) patients surviving for more than 4 years. In this case, disease progression was elicited 9 months after surgery, and the patient survived for 18 months.

In patients with gliomas, Ommaya reservoir provides a clinical improvement when used for recurrent aspiration in patients with cystic tumors.^[1] It may be used as a temporary measure if complementary treatment or re-intervention is being administered or planned. It can also be used for

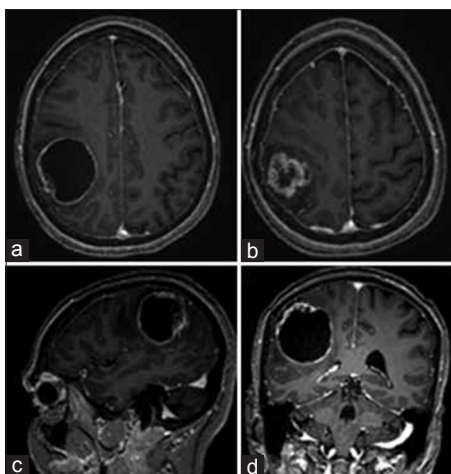


Figure 1: Head magnetic resonance imaging. (a and b) Axial T1 gadolinium; (c) sagittal T1 gadolinium; (d) coronal T1 gadolinium: Right subcortical frontoparietal lesion with cystic and solid components with enhancement after contrast administration, vasogenic edema and mass effect with partial collapse of the right lateral ventricle

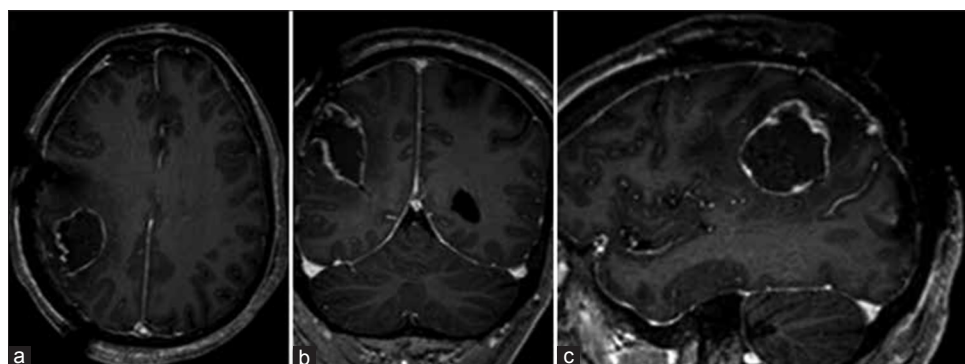


Figure 2: Postoperative head magnetic resonance imaging: (a) Axial T1 gadolinium; (b) coronal T1 gadolinium; (c) sagittal T1 gadolinium: Right subcortical frontoparietal mass lesion with decreased mass effect and mild cystic enhancement after subtotal removal

intracystic brachytherapy which might have been an option if no hemorrhagic event had happened. Although the

clinical impact of such approach is unknown, the literature supports some benefit when used in a compassionate add-on treatment.^[19]

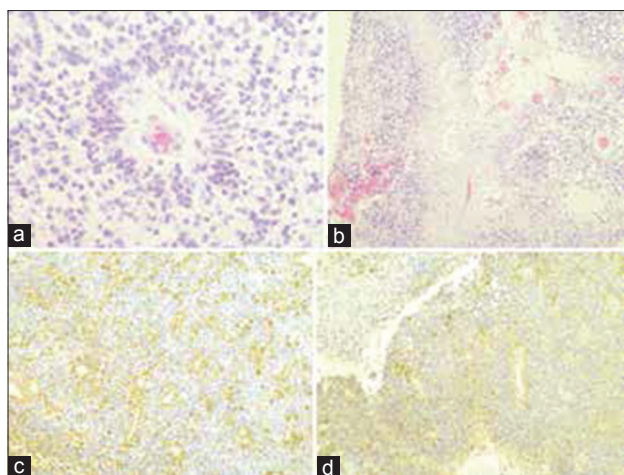


Figure 3: Glial neoplasm whose elements were predominantly arranged in vascular pseudo-rosettes. (a) Mitotic index was brisk, and there were microvascular proliferation and areas of necrosis. (b) Immunohistochemical study showed mild and focal glial fibrillary acidic protein (c) and epithelial membrane antigen (d) but strong S-100 protein and vimentin immunoreactivity. (a) - Neoplastic elements arranged in vascular pseudo-rosettes (H and E $\times 40$); (b) necrotic areas of the tumor (H and E $\times 10$); (c) neoplastic elements glial fibrillary acidic protein immunoreactives ($\times 10$); (d) neoplastic elements epithelial membrane antigen immunoreactives ($\times 10$)

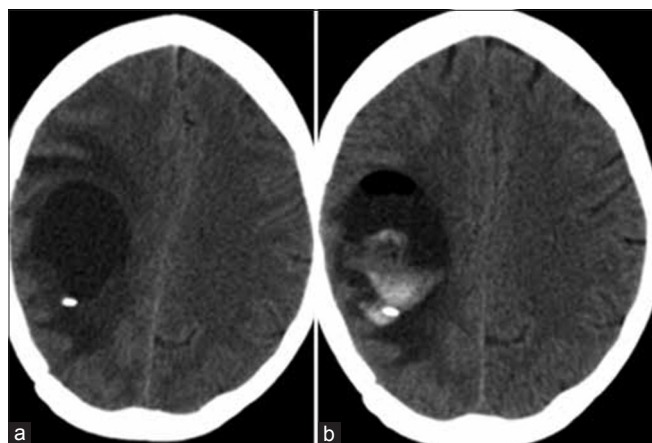


Figure 4: Head computed tomography: (a and b) (Axial noncontrast): Intracystic hemorrhage after Ommaya reservoir drainage. Arrow – Ommaya reservoir catheter tip

The literature considers as poor prognostic factors: Young age, incomplete tumor resection, histological anaplasia, supratentorial, and extraventricular location.^[6,11] By definition, SEAE comprises the last three, leaving age and extension of surgical resection as important prognostic factors in these patients. We consider tumor location in eloquent areas is an important limiting factor for a complete surgical resection which is the main modifiable negative prognostic factor in SEAE. Complete resection should be attempted in all cases, and reintervention should be considered on a case basis when first surgery limiting factors are overcome.

To our best knowledge, this is the first report where an Ommaya reservoir was used for symptom control while waiting for adjuvant treatment to control cystic fluid production after incomplete resection in an eloquent area.

Conclusion

Adult SEAE is a rare entity raising controversy regarding its best approach. Some poor prognostic features are identified: Young age and incomplete tumor resection. Tumor location in the eloquent area is also a factor of poor outcome as it usually precludes complete resection. Ommaya reservoirs may be used in cystic lesions as a temporary measure. Gross total resection is the preferred approach followed by adjuvant treatment with radiotherapy (no consensual place for chemotherapy).

Patient consent

The next of kin has consented to the submission of the case report for submission to the journal.

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Nil.

Conflicts of interest

There are no conflicts of interest.

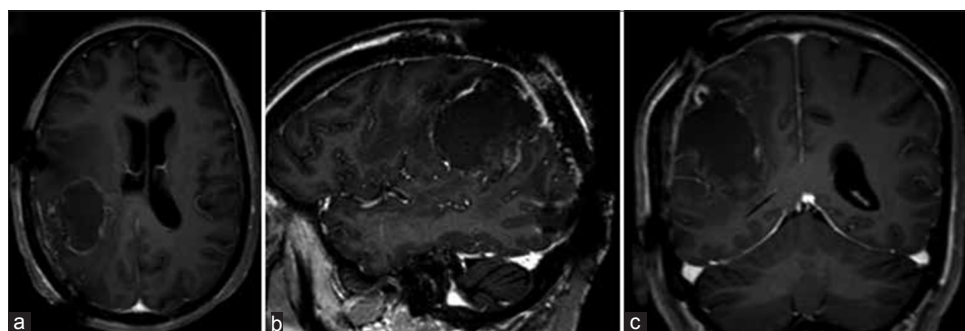


Figure 5: Second surgery postoperative head magnetic resonance imaging. (a) (Axial T1 gadolinium), (b) (sagittal T1 gadolinium), and (c) (coronal T1 gadolinium): Right subcortical frontoparietal lesion with cystic component and minimal ring enhancement after contrast administration - less contrast enhancement when compared with the first surgery postoperative Head magnetic resonance imaging [Figure 2]

Table 1: Clinical review of published SEAE

Authors; year of publication	Gender, age (years)	Symptoms	Localization, size, features	Treatment	Follow-up; survival period
Takeshima <i>et al.</i> , 2002 ^[16]	Female, 70	Consciousness disturbance and head trauma; progressive dementia	Right frontal lobe, intratumoral hemorrhage	Macroscopic total resection; second operation due to hemorrhage; bedridden	No recurrence for 14 months
Moritani <i>et al.</i> , 2003 ^[13]	Female, 50	Severe headache	Right temporal lobe, 20 mm	Macroscopic total resection; chemotherapy	Recurrence 7 and 13 months after first surgery with reoperation and chemotherapy and irradiation after the first recurrence. Survival for 20 months
Kojima <i>et al.</i> , 2003 ^[12]	Female, 56	Transient mild vertigo, seizure of the right extremities, mild dysarthria, agraphia, headache	Left temporal lobe, hematoma, calcification	Subtotal resection; focal radiation therapy	
Shuangshoti <i>et al.</i> , 2005 ^[15]			Left parietal	Surgery; radiation; chemotherapy	No recurrence; survival for 18 years
			Right frontal		Unknown; survival for 14 years
			Left parietal	Surgery; radiation	No recurrence; survival for 2 years
			Left frontal		Unknown; survival for 4, 8 years
			Right occipital	Surgery; radiation	No recurrence; survival for 5 years
			Frontal	Surgery; radiation; chemotherapy	Recurrence after 3 years
Miyazawa <i>et al.</i> , 2007 ^[5]	Male, 33	Acute headache, vomiting, transient motor aphasia	Right frontal Left angular gyrus; intratumoral hemorrhage; calcification; adhesion to the dura mater	Total macroscopic resection; IAR (interferon-B and ACNU, local irradiation 60 Gy)	Unknown; survival for 14 months Recurrence 6 months later
Niazi <i>et al.</i> , 2009 ^[4]	Male, 18	Simple partial motor seizures for 5 months	Right frontal; 4 cm	Gross total resection; radiation	Recurrence; death 14 months after
	Female, 36	Generalized clonic-tonic seizure; headaches for 3 months	Right frontal lobe; 2,4 cm	Gross total resection; radiation	No recurrence; uneventful for 29 months
Davis <i>et al.</i> , 2011 ^[9]	Female, 22	Headache and dysarthria	Frontotemporal	Macroscopic total resection; radiotherapy	Metastatic systemic disease 1 year after treatment; 5 years follow-up
Romero <i>et al.</i> , 2012 ^[14]	Male, 23	Seizures, hemiparesis, and aphasia	Frontal lobe	Complete macroscopic removal; radiotherapy	No recurrence during 5 years of follow-up
Elsharkawy <i>et al.</i> , 2013 ^[10]	Male, 25	Seizure, finger paresthesia	Frontoparietal lesion	Complete macroscopic removal; radiotherapy	Follow-up of 6 months with no recurrence
Iwamoto <i>et al.</i> , 2014 ^[11]	Male, 61	Severe acute headache for 3 days, consciousness disturbance, coma	Right temporal lobe, 40 mm, repeated intratumoral hemorrhage	Macroscopic total resection; focal radiation therapy (60 Gy) and chemotherapy (temozolomide)	Recurrence and dissemination to lower thoracic spinal cord

SEAE – Supratentorial extraventricular anaplastic ependymoma; IAR – ACNU-Radiation therapy; ACNU – 1-(4-amino-2-methyl-5-pyrimidinyl)-methyl-3-(2-chloroethyl)-3-nitrosourea hydrochloride

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