Case Report

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 infratentorially, develops 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 cvst wall with no anatomical cleavage plan, no gross total resection was possible. An Ommava reservoir was inserted. Postoperative MRI showed mild enhancement of the cystic wall and decreased mass effect [Figure 2]. pathological diagnosis The 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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José Pedro Lavrador, Edson Oliveira, Joaquim Cruz Teixeira, José Pedro Lopes, José Pimentel¹, Manuel Herculano Carvalho

Department of Neurosurgical, Hospital Santa Maria, Centro Hospitalar Lisboa Norte, Lisbon, 'Neuropathology Laboratory, Hospital Santa Maria, Centro Hospitalar Lisboa Norte, Lisbon, Portugal

Address for correspondence: Dr. José Pedro Lavrador, Rua Cidade de Faro, N° 40, 2725-689 Mem-Martins, Lisboa, Portugal. E-mail: jose.pedro.lavrador@ gmail.com



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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.

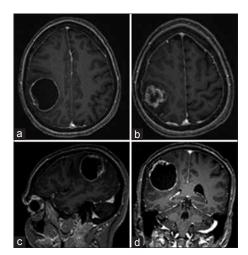


Figure 1: Head magnetic resonance imaging. (a and b) Axial T1 gadolinium; (c) sagittal T1 gadolinium; (d) coronal T1 gadolinium: Right subcortical frontoparietal lesion with cystic and solid components with enhancement after contrast administration, vasogenic edema and mass effect with partial collapse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

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 cvst 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

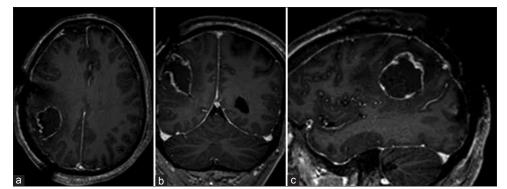


Figure 2: Postoperative head magnetic resonance imaging: (a) Axial T1 gadolinium; (b) coronal T1 gadolinium; (c) sagittal T1 gadolinium: Right subcortical frontoparietal mass lesion with decreased mass effect and mild cystic enhancement after subtotal removal

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

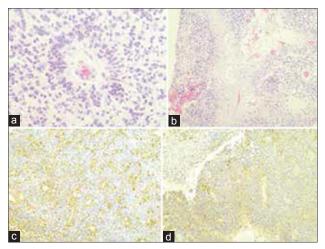


Figure 3: Glial neoplasm whose elements were predominantly arranged in vascular pseudo-rosettes. (a) Mitotic index was brisk, and there were microvascular proliferation and areas of necrosis. (b) Immunohistochemical study showed mild and focal glial fibrillary acidic protein (c) and epithelial membrane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

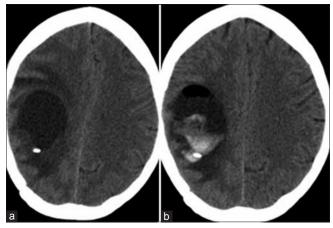


Figure 4: Head computed tomography: (a and b) (Axial noncontrast): Intracystic hemorrhage after Ommaya reservoir drainage. Arrow – Ommaya reservoir catheter tip

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

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 approached 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.

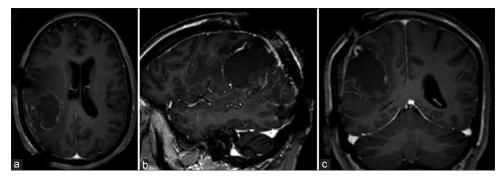


Figure 5: Second surgery postoperative head magnetic resonance imaging. (a) (Axial T1 gadolinium), (b) (sagittal T1 gadolinium), and (c) (coronal T1 gadolinium): Right subcortical frontoparietal lesion with cystic component and minimal ring enhancement after contrast administration - less contrast enhancement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

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Authors; year of	Gender,	Symptoms	Localization, size, features	Treatment	Follow-up; survival period
publication	age (years)		leatures		
Takeshima	Female,	Consciousness	Right frontal	Macroscopic total	No recurrence for 14 months
et al., 2002 ^[16]	70	disturbance and head	lobe, intratumoral	resection; second	
		trauma; progressive	hemorrhage	operation due to	
		dementia		hemorrhage; bedridden	
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	Female,	Transient mild	Left temporal	Subtotal resection; focal	
et al., 2003 ^[12]	56	vertigo, seizure of the right extremities, mild dysarthria, agraphia, headache	lobe, hematoma, calcification	radiation therapy	
Shuangshoti et al., 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 Frontal	Surgery; radiation Surgery; radiation; chemotherapy	No recurrence; survival for 5 years Recurrence after 3 years
			Right frontal		Unknown; survival for 14 months
Miyazawa et al., 2007 ^[5]	Male, 33	Acute headache, vomiting, transient motor aphasia	Left angular gyrus; intratumoral hemorrhage; calcification; adhesion to the dura mater	Total macroscopic resection; IAR (interferon-B and ACNU, local irradiation 60 Gy)	Recurrence 6 months later
Niazi et al., 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 et al., 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 et al., 2013 ^[10]	Male, 25	Seizure, finger paresthesia	Frontoparietal lesion	Complete macroscopic removal; radiotherapy	Follow-up of 6 months with no recurrence
Iwamoto et al., 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-cholroethyl)-3-nitrosourea hydrochloride

References

- Amirian ES, Armstrong TS, Gilbert MR, Scheurer ME. Predictors of survival among older adults with ependymoma. J Neurooncol 2012;107:183-9.
- 2. Mermuys K, Jeuris W, Vanhoenacker PK, Van Hoe L,

D'Haenens P. Best cases from the AFIP: Supratentorial ependymoma. Radiographics 2005;25:486-90.

3. Metellus P, Barrie M, Figarella-Branger D, Chinot O, Giorgi R, Gouvernet J, *et al.* Multicentric French study on adult intracranial ependymomas: Prognostic factors analysis and therapeutic considerations from a cohort of 152 patients. Brain

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2007;130(Pt 5):1338-49.

- Niazi TN, Jensen EM, Jensen RL. WHO Grade II and III supratentorial hemispheric ependymomas in adults: Case series and review of treatment options. J Neurooncol 2009;91:323-8.
- Miyazawa T, Hirose T, Nakanishi K, Uozumi Y, Tsuzuki N, Shima K. Supratentorial ectopic cortical ependymoma occurring with intratumoral hemorrhage. Brain Tumor Pathol 2007;24:35-40.
- Roncaroli F, Consales A, Fioravanti A, Cenacchi G. Supratentorial cortical ependymoma: Report of three cases. Neurosurgery 2005;57:E192.
- Vernet O, Farmer JP, Meagher-Villemure K, Montes JL. Supratentorial ectopic ependymoma. Can J Neurol Sci 1995;22:316-9.
- Antony R, Wong KE, Patel M, Olch AJ, McComb G, Krieger M, *et al.* A retrospective analysis of recurrent intracranial ependymoma. Pediatr Blood Cancer 2014;61:1195-201.
- Davis MJ, Hasan F, Weinreb I, Wallace MC, Kiehl TR. Extraventricular anaplastic ependymoma with metastasis to scalp and neck. J Neurooncol 2011;104:599-604.
- Elsharkawy AE, Abuamona R, Bergmann M, Salem S, Gafumbegete E, Röttger E. Cortical anaplastic ependymoma with significant desmoplasia: A case report and literature review. Case Rep Oncol Med 2013;2013:354873.
- Iwamoto N, Murai Y, Yamamoto Y, Adachi K, Teramoto A. Supratentorial extraventricular anaplastic ependymoma in an adult with repeated intratumoral hemorrhage. Brain Tumor Pathol 2014;31:138-43.
- 12. Kojima A, Yamaguchi N, Okui S, Kamiya M, Hirato J, Nakazato Y. Parenchymal anaplastic ependymoma with

intratumoral hemorrhage: A case report. Brain Tumor Pathol 2003;20:85-8.

- Moritani S, Kushima R, Bamba M, Kobayashi TK, Oka H, Fujimoto M, *et al.* Highly anaplastic extraventricular ependymoma arising in an adult, mimicking metastatic adenocarcinoma with heavy stromal inflammation and emperiporesis. Pathol Int 2003;53:539-46.
- Romero FR, Zanini MA, Ducati LG, Vital RB, de Lima Neto NM, Gabarra RC. Purely cortical anaplastic ependymoma. Case Rep Oncol Med 2012;2012:541431.
- Shuangshoti S, Rushing EJ, Mena H, Olsen C, Sandberg GD. Supratentorial extraventricular ependymal neoplasms: A clinicopathologic study of 32 patients. Cancer 2005;103:2598-605.
- Takeshima H, Kawahara T, Uchida H, Hirano H, Nakazato Y, Kuratsu J. Brain surface ependymoma with repeated episodes of intratumoral hemorrhage – Case report. Neurol Med Chir (Tokyo) 2002;42:166-9.
- Borkar SA, Subbarao KC, Sharma MC, Mahapatra AK. Cystic with mural nodule: Unusual radiological presentation of supratentorial anaplastic ependymoma. J Pediatr Neurosci 2012;7:101-2.
- Furie DM, Provenzale JM. Supratentorial ependymomas and subependymomas: CT and MR appearance. J Comput Assist Tomogr 1995;19:518-26.
- Musolino A, Merckaert P, Munari C, Daumas-Duport C, Chodkiewicz JP. Stereotactic endocavitary treatment of cysts and pseudocysts of glioma. Preliminary report. J Neurosurg Sci 1989;33:107-14.