Case Report

ANNALS OF MEDICINE & SURGERY

OPEN

Supratentorial extra-axial anaplastic ependymoma: a rare case report

Abukar Mahamed Osman, MD[a,b,*], Ulvi Çiftçi, MD[b], Nurten Sever, MD[b]

Introduction and importance: Ependymomas are central nervous system tumors arising from the ependymal lining of the ventricle and spinal cord. Supratentorial extra-axial ependymomas are very rare, most commonly affecting the pediatric population and rarely in adults.

Case presentation: The authors report a case of a 71-year-old female with a headache and blurred vision. An MRI scan revealed a lesion at the parafalcine region of the occipital lobe. A parieto-occipital craniotomy was performed. When the dura was opened during the operation, the extra-axially located, well-circumscribed, dirty yellow-white tumor dissected from the surrounding tissue was excised entirely by microdissection. Histopathological examination revealed supratentorial extra-axial anaplastic ependymoma. The patient received postoperative radiation therapy (54 Gray over 30 fractions). No recurrence of the tumor was observed during the 4-year follow-up.

Discussion: Supratentorial ependymomas at the extra-axial region are uncommon; extra-axial anaplastic ependymoma and meningiomas have similar radiological findings, such as a dural tail, subarachnoid plane, and diffuse enhancement after contrast injection. This close similarity might cause misdiagnoses. Total surgical resection was followed by adjuvant radiotherapy and close follow-up in the gold standard of the treatment.

Conclusion: The authors report a rare case of anaplastic ependymomas located at the extra-axial region. Anaplastic tumors are prone to recurrence despite total resection and radiation therapy; hence, a close follow-up is warranted.

Keywords: anaplastic, ependymoma, extra-axial, supratentorial

Introduction

Ependymomas are central nervous system tumors arising from the ependymal lining of the ventricle and spinal cord. They account for about 2% of all intracranial tumors, most commonly affecting the pediatric population. In adults, they represent 3–5% of glial tumors[1,2].

Most ependymomas develop infratentorial, and they may also arise at the supratentorial and mainly near the walls of the ventricles, consistent with the location of the ependymal cells from which they originate[3].

Approximately, half of the supratentorial ependymomas are extraventricular, while the 'pure' extra-axial ependymomas with no connection to the cortex are rare. Additionally, anaplastic ependymomas localized in this region are rare[4,5]. Radiologically, these lesions can be confused with meningioma preoperatively because of their appearance, similar to convexity meningioma[6,7].

Although this will not change the treatment strategy in the first step, it is necessary to investigate anaplastic ependymomas further for seeding metastasis. Therefore, histopathological examination becomes imperative for proper evaluation and an adequate diagnosis. This case report has been written in accordance with Surgical CAse REport (SCARE) guidelines[8].

HIGHLIGHTS

- Supratentorial extra-axial ependymomas are very rare, most commonly affecting the pediatric population and rarely in adults.
- In previous cases in the literature, all were younger than our case, and our patient is the oldest case ever reported.
- A total resection should be the goal of the surgery, and postoperative radiotherapy is recommended, anaplastic tumors are prone to recurrence despite total resection and radiation therapy; hence, a close follow-up is warranted.

Case report

The present case is a 71-year-old female suffering from blurred vision and headache for 1 month. Her neurologic examination

[a]Mogadishu Somalia Turkish Training and Research Hospital, Mogadishu, Somalia and [b]Istanbul Bakirkoy Dr. Sadi Konuk Training and Research Hospital, Istanbul, Turkey

Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

*Corresponding author. Address: Wadajir District, Mogadishu, Somalia. Tel.: +252 615246933. E-mail: abukarmcj@gmail.com (A.M. Osman).

Annals of Medicine & Surgery (2023) 85:5634–5640

Received 17 February 2023; Accepted 18 March 2023

Published online 5 September 2023

http://dx.doi.org/10.1097/MS9.0000000000000536

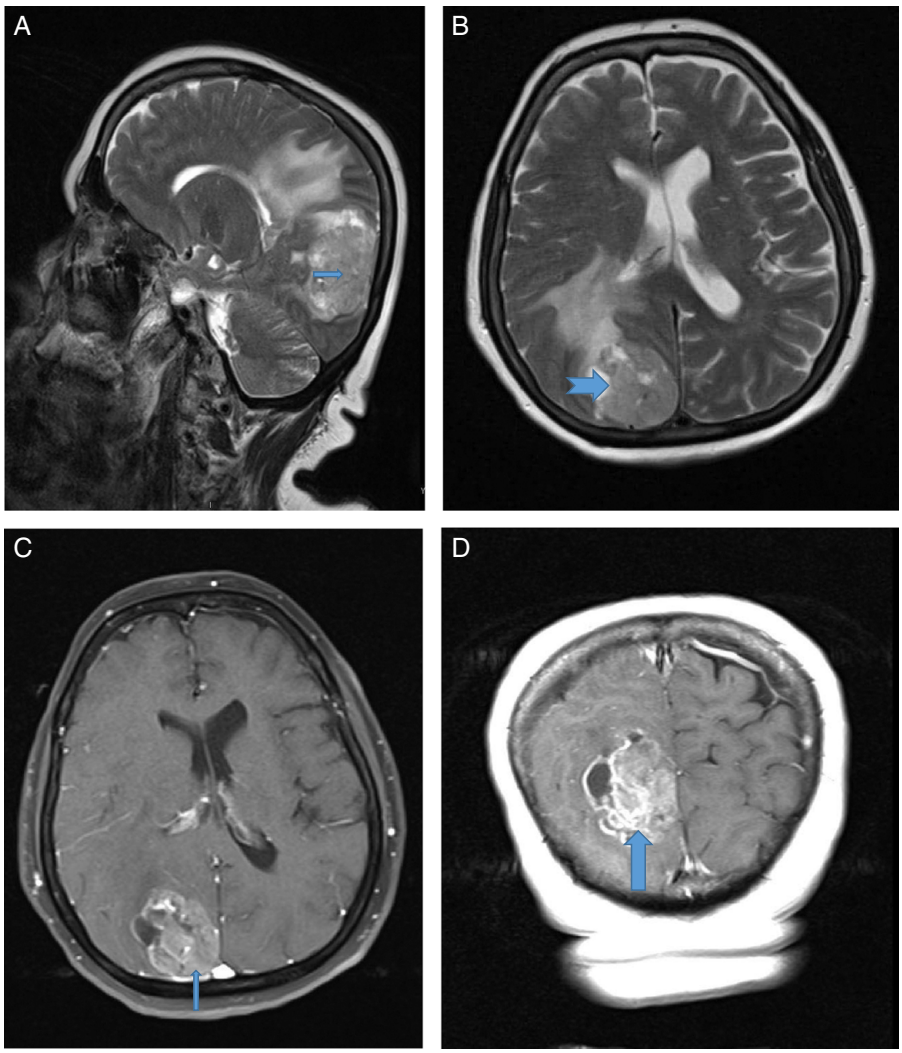

Figure 1. Preoperative: A T2 -weighted image showed hyperintense signal characteristics due to cystic components observed in the lesion. (A) An axial T2-weighted image; (B) a sagittal T2 image; (C, D) gadolinium-enhanced T1 – at the level of the right occipital lobe, a mass lesion of ~40 × 38 mm with heterogeneous contrast enhancement is observed in the axial plane and coronal image.

was intact. MRI revealed a lesion at the parafalcine region of the occipital lobe. It showed an isointense signal in T1 and T2 with extensive parenchymal edema; the contrast study showed heterogeneous enhancement (Fig. 1). The patient was admitted to the neurosurgery clinic for an operation.

A parieto-occipital craniotomy was performed. When the dura was opened during the operation, the extra-axially located, well-circumscribed, dirty yellow-white tumor was easily dissected from the surrounding tissue. The tumor was excised entirely by microdissection. There were no intraoperative and postoperative complications documented. Histopathological examination revealed a glial tumor with focal infiltration and an extensive, expansive growth pattern in the brain parenchyma (supratentorial extra-axial anaplastic ependymoma (EAAE).

Dystrophic calcification and hypercellularity, pleomorphism, mitotic activity, vascular endothelial proliferation, and necrosis were detected in places, consisting of cells with fine chromatin and prominent nucleoli, with the perivascular arrangement, and partly perinuclear halo, partly irregular nuclear contour (Figs 2 and 3). The Ki-67 proliferation index was 20–30%. The pathology report came as 'clear cell anaplastic ependymoma' (WHO grade III). No metastasis was found in additional radiological imaging in terms of seeding metastasis. The patient received postoperative radiation therapy (54 Gray over 30 fractions).

At 6-month follow-up, the patient's clinical condition subsided utterly, and a contrast magnetic resonance (MRI) showed no evidence of residual tumor (Fig. 4). Follow-ups of 1 year, 2 years (Figs 5 and 6) and 4 years reported no tumor recurrence.

Discussion

Ependymomas are tumors of the central nervous system that originate from ependymal cells. These cells are mostly known as the cell type lining the ventricles of the brain and the central canal of the spinal cord[9]. Although most pediatric patients show an

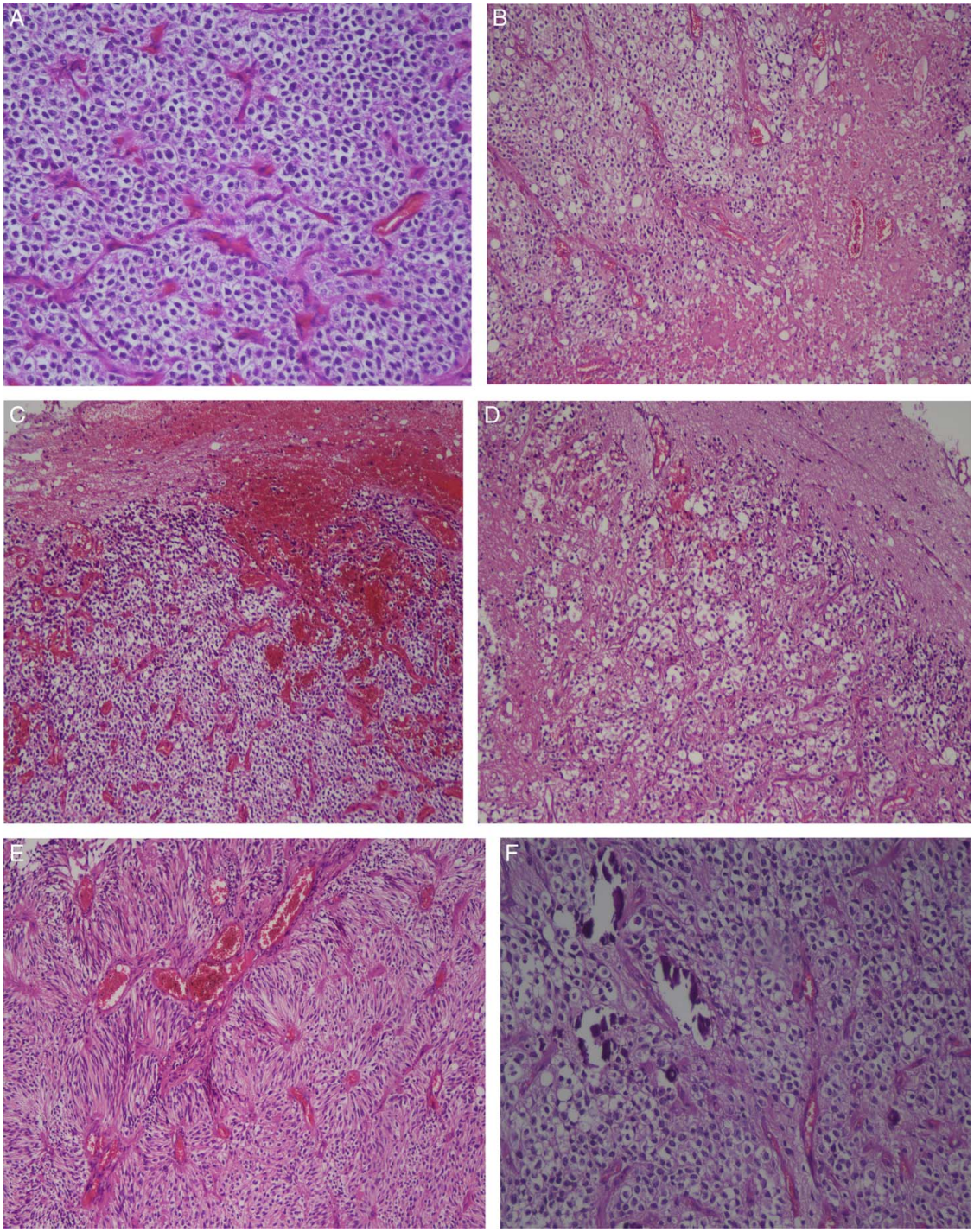

Figure 2. Histopathologic examination of hematoxylin and eosin (H&E) stained slides showed a cellular tumor arranged in nodules and sheets with delicate capillary blood vessels. (A) Neoplastic cells with perinuclear halo, arranged around prominent vascular roof H + E × 100. (B) Tumor necrosis. (C) Glial tumor with expansive growth pattern H + E × 40. (E) Tumor with expansive growth pattern H + E × 100. (D) Cells with narrow extension cytoplasm forming pseudorosette H + E × 100. (E) Calcifications foci H + E × 100.

infratentorial location, a supratentorial predominance is present in the adult population[10]. Supratentorial ependymomas, located at the extra-axial region, are rare; moreover, EAAE is extremely rare. In the literature, only 14 cases of anaplastic ependymomas located at the extra-axial region have been reported previously[5,11–22] (Table 1).

EAAE and meningiomas have similar radiological findings, such as a dural tail, subarachnoid plane, and diffuse enhancement after contrast injection. This close similarity might cause misdiagnoses. This situation may be acceptable at the first step in the treatment. The aim of the treatment for both EAAE and meningioma is a total surgical excision. However, the clinical course of

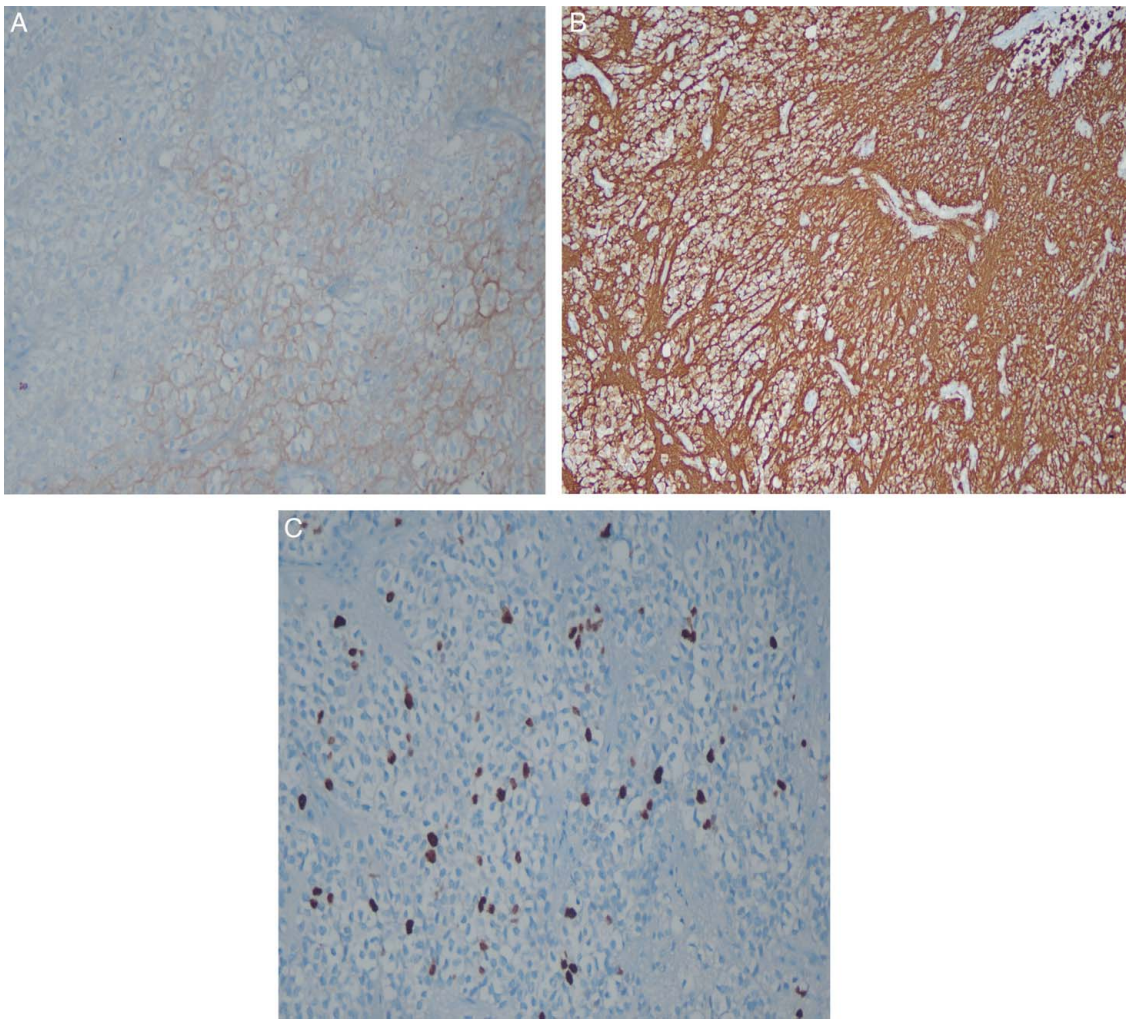

Figure 3. On immunohistochemistry, the tumor cells showed perivascular dot-like positivity for : EMA (×100) (A), strong immunopositivity for (B), GFAP with perivascular accentuation (×100) (C), and high Ki-67 (×100).

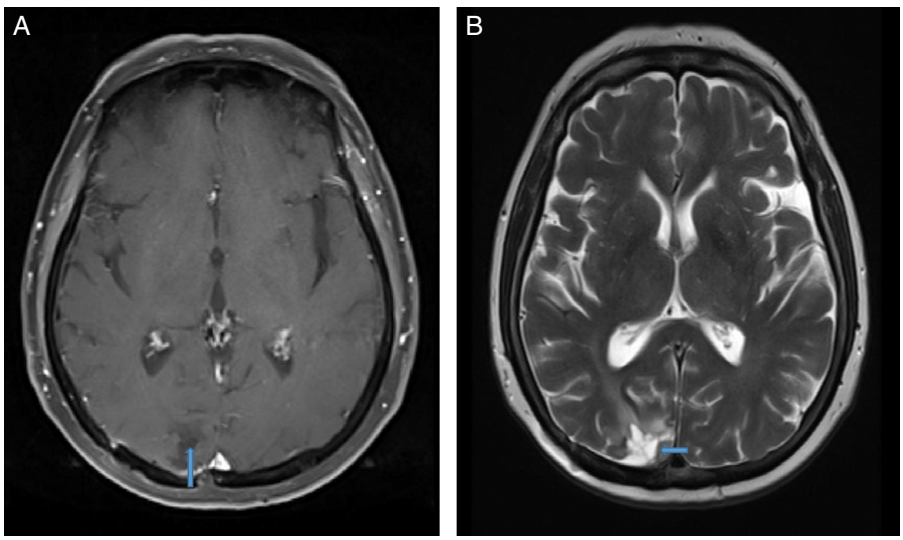

Figure 4. Postoperative 6-month. (A) Axial T1 image, there is no gadolinium-enhanced. (B) Axial T2 image, there is hyperintensity (encephalomalacia) tissue loss of the brain parenchyma in the right occipital lobe section of the patient with a previous operation history.

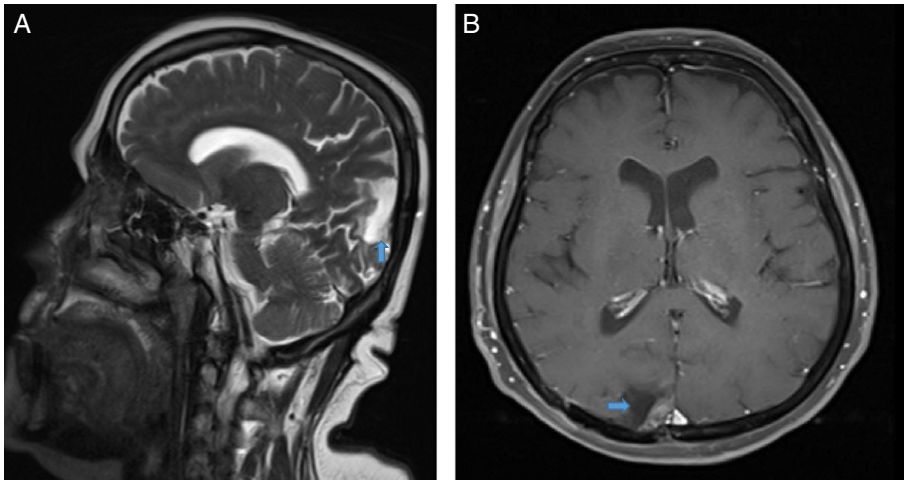

Figure 5. Postoperative 1 year. In the right occipito-parietal region, there is an area of sequela cystic encephalomalacia that includes the cortex-subcortical white matter in the brain parenchyma. (A) Sagittal T2 image; (B) T1 axial image with i.v. contrast.

the EAAE is different from meningioma due to the possibility of local recurrence and seeding metastasis. For this reason, close follow-up for the postoperative histologic diagnosis is crucial.

In previous cases in the literature, all were younger than our case, and our patient is the oldest case ever reported.

Six patients' seizures were the main presenting symptoms[5,11,14,15,19,20] (Table 1). Thus, it can be related to a natural result of a space-occupying mass that irritates the cortex. In the case of a slowly progressing meningioma, the seizure would not be a clinically highly anticipated finding. This symptom may be a distinctive feature of the EAAE from meningiomas. Our patient's complaints were blurred vision and headache, which differ from the presenting symptoms of the other cases in the literature.

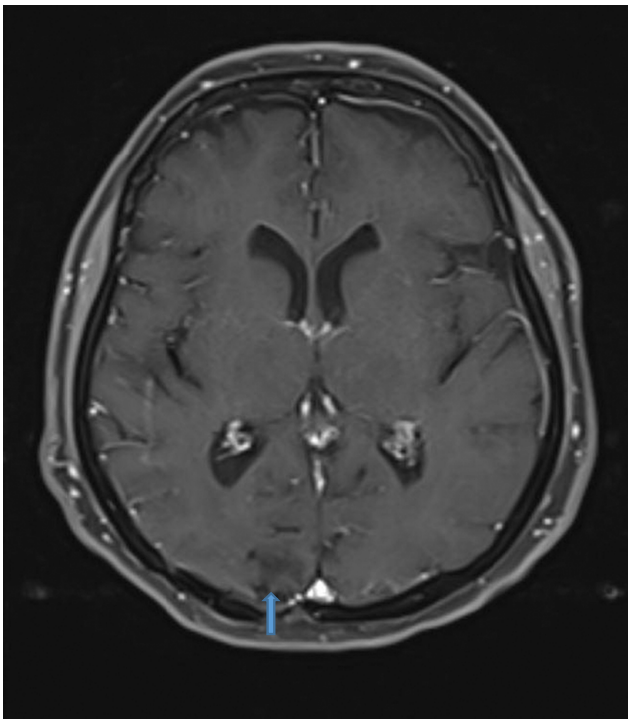

Figure 6. Postoperative 2 year. No contrast enhancement is observed in the axial T1 image, the right posterior parietal region adjacent to the encephalomalacia area.

Considering the radiological features of the reported cases, it seems that EAAE does not have a radiological feature that clearly distinguishes it from meningioma.

Singh *et al.*[11] mention that meningiomas frequently have a dural tail. Nevertheless, as a feature of extra-axial localization, most cases appear to have subarachnoid planes. It is noteworthy that features such as the subarachnoid plane, the dural tail, and the dural attachment are sporadic.

Several hypotheses have been proposed, explaining the appearance of ependymomas in an unexpected area, such as the extra-axial area. Among these hypotheses, the most favored one is the thought that the tumor originated from ectopic ependymal nests resulting from migration errors of the germinal matrix[23].

Surgical resection remains the gold standard for the management of these tumors. A systematic review regarding the optimal treatment option for supratentorial ependymomas in adults concluded that all WHO grade III ependymomas require adjuvant radiotherapy. In contrast, total resection without radiotherapy can manage all WHO grade II ependymomas[24]. The extent of the surgical removal of the mass remains the most important prognostic factor. Mansur *et al.*[25] reported a recurrence rate in up to half of the patients with WHO grade III ependymoma. Despite our case being elderly and having a grade III anaplastic ependymoma, total surgical resection was followed by adjuvant radiotherapy. For a 4-year follow-up, no recurrence was detected.

Conclusion

We report a rare case of anaplastic ependymomas located at the supratentorial extra-axial region in the oldest patient in the literature. A total resection should be the goal of the surgery, and postoperative radiotherapy is recommended. Moreover, the patient must be evaluated further for seeding metastasis and postoperative histologic diagnosis. Anaplastic tumors are prone to recurrence despite total resection and radiation therapy; hence, a close follow-up is warranted.

Table 1

Literature review of the previously reported cases including our patient.

References	Age/ sex	Location/preoperative tumor size (cm)	Symptoms	Preoperative diagnosis (intraoperative diagnosis)	Surgery	Histology/WHO grade	Adjuvant treatment	follow-up period
Hanchery *et al.*[12]	29/ M	Interhemispheric/ 6.5 × 5.5 × 7.5	Headache	Meningioma,	TR	Classical Ep/II	RT, CT	No follow-up
Hayashi *et al.*[13]	13/M	Rt occipital-parietal/NA	Headache	Cystic meningioma Meningeal sarcoma	GTR	Clear-cell Ep/II	None	3 m NR
Youkilis *et al.*[14]	20/male	Lt parafalcine/ 5.0 × 3.0 × 3.2	Seizure	Meningioma	GTR	Clear-cell Ep/III	None	12 m NR
Park *et al.*[15]	17/female	R- frontoparietal area 7 × 4 cm	Seizure	Meningioma	GTR		RT	No
Salunke *et al.*[16]	43, F	Lt posterior one-third parasagittal/7.2 × 5.9 × 4	loss of vision and headache	Meningioma (meningioma, favor)	GTR	Classical Ep/II	RT	6 m NR
Sigh *et al.*[11]	35/male	Lt middle-third parafalcine (1.8 cm)	Seizures	Meningioma	GTR	Anaplastic ED* WHO Grade III	RT	12 m NR
Dilli *et al.*[17]	15/female	Lt frontoparietal 6.5 × 3.5	Syncope	not defined	GTR	Anaplastic ED* WHO Grade III.	RT	not mentioned
Nambirajan *et al.*[18]	9/ F	Rt frontal parafalcine/NA	Headache	Meningioma Hemangiopericytoma	GTR	Anaplastic, focal clear-cell, Ep/III	RT	6 m NR
Gupta *et al.*[5]	9/male	Rt frontoparietal area 6.4 × 5.2 cm	Seizures	Meningioma	GTR	Anaplastic, Ep/WHO III	RT	15 m NR
Yang *et al.*[19]	47, M	Lt middle-third parafalcine/ 3.8 × 3.2 × 2.1	Seizure	Meningioma	GTR	Anaplastic Ep/III	RT	53 m NR
Yang *et al.*[19]	30, M	Rt temporal/3.8 × 3.0 × 3.5 Rt occipital/6.0 × 5.6 × 6.5	Seizure	Glioma Glioma	NTR GTR GTR	Anaplastic Ep/III Anaplastic Ep/III	None RT RT	3–5 m R 28 m NR 26 m NR
Satyarthee *et al.*[20]	9, F	Rt middle-third parafalcine/ 8.6 × 6.0 × 5.4	Seizure	Meningioma	GTR	Anaplastic Ep/III	RT	16 m NR
Karthigeyan *et al.*[21]	33, F	Rt front-parietal/NA	headache	Meningioma Gliosarcoma Hemangiopericytoma	GTR	Anaplastic Ep/III	RT	12 m NR
Nagatasu *et al.*[22]	26, M	Lt parietal-temporal/ 7.0 × 5.3 × 5.7	headache	Meningioma (meningioma)	NTR GTR	Anaplastic Ep/III	RT	12 m R 48 m R 52 DOD
Present case	71/F	parafalcine region of the occipital lobe	headache blurred vision	not defined	GTR	Anaplastic Ep/III	RT	48 m NR

CT, chemotherapy; DOD, death of disease; Ep, ependymoma; F, female; GTR, gross total resection; Lt, left; M, male; NA, not available; NR, no recurrence; NTR, near total resection; R, recurrence; RT, radiotherapy; Rt, right; TR, total resection.

Ethical approval

Ethical approval was not needed for writing a case report in our settings.

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Sources of funding

No sources of funding.

Author contribution

A.M.O.: concept design and preparation of the manuscript; U.Ç.: literature search; A.M.O., U.Ç., and N.S.: preparation of draft manuscript.

Conflicts of interest disclosure

All authors declare that they have no conflicts of interest.

Research registration unique identifying number (UIN)

1. Name of the registry: not applicable.
2. Unique identifying number or registration ID: not applicable.
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): not applicable.

Guarantor

Abukar Mahamed Osman.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Study design

Case report.

References

[1] Lavrador J, Oliveira E, Teixeira J, *et al*. Adult supratentorial extraventricular anaplastic ependymoma: therapeutic approach and clinical review. Asian J Neurosurg 2018;13:105–9.

[2] Wang M, Zhang R, Liu X, *et al*. Supratentorial cortical ependymomas: a retrospective series of 13 cases at a single center. World Neurosurg 2018; 112:e772–7.

[3] Leng X, Tan X, Zhang C, *et al*. Magnetic resonance imaging findings of extraventricular anaplastic ependymoma: a report of 11 cases. Oncol Lett 2016;12:2048–54.

[4] Schwartz TH, Kim S, Glick RS, *et al*. Supratentorial ependymomas in adult patients. Neurosurgery 1999;44:721–31.

[5] Gupta VK, Kapoor I, Radotra BD, *et al*. Pure extra-axial supratentorial anaplastic ependymoma with no cortical attachment: a rare case. Neurol India 2016;64:345.

[6] Ahn S, Kim YJ, Lee YS, *et al*. Extra-axial and clear cell type ependymoma , mimicking a convexity meningioma. Brain Tumor Res Treat 2017;5: 127–30.

[7] Ahn S, Kim YJ, Lee YS, *et al*. Extra-axial and clear cell type ependymoma, mimicking a convexity meningioma. Brain Tumor Res Treat 2017;5: 127–30.

[8] Agha RA, Franchi T, Sohrabi C, *et al*. The SCARE 2020 guideline: updating consensus Surgical Case REport (SCARE) guidelines. Int J Surg 2020;84:226–30.

[9] Gerstner ER, Pajtler KW. Ependymoma. InSeminars Neurol 2018;38 (No. 01):104–11; Thieme Medical Publishers.

[10] Mermuys K, Jeuris W, Vanhoenacker PK, *et al*. Supratentorial ependymoma. Radiographics 2005;25:486–90.

[11] Singh V, Turel M, Chacko G, *et al*. Supratentorial extra-axial anaplastic ependymoma mimicking a meningioma. Neurol India 2012;60:111.

[12] Hanchey RE, Stears JC, Lehman RA, *et al*. Interhemispheric ependymoma mimicking falx meningioma: case report. J Neurosurg 1976;45: 108–12.

[13] Hayashi K, Tamura M, Shimozuru T, *et al*. Extra-axial Ependymoma—Case Report—. Neurol Med Chir (Tokyo) 1994;34:295–9.

[14] Youkilis AS, Park P, McKeever PE, *et al*. Parasagittal ependymoma resembling falcine meningioma. AJNR Am J Neuroradiol 2001;22: 1105–8.

[15] Park EK, Lee YH, Kim DS, *et al*. 17-year-old girl with headache and complex partial seizure. Brain Pathol 2010;20:1111.

[16] Salunke P, Kovai P, Sura S, *et al*. Extra-axial ependymoma mimicking a parasagittal meningioma. J Clin Neurosci 2011;18:418–20.

[17] Dilli UD, Yıldırım M, Süren D, *et al*. Frontal lobe ependymoma: a case report. Erciyes Med J/Erciyes Tip Dergisi 2013;35:75–8.

[18] Nambirajan A, Malgulwar PB, Sharma MC, *et al*. C11orf95-RELA fusion present in a primary intracranial extra-axial ependymoma: report of a case with literature review. Neuropathology 2016;36:490–5.

[19] Yang Y, Tian KB, Hao SY, *et al*. Primary intracranial extra-axial anaplastic ependymomas. World Neurosurg 2016;90:704–e1.

[20] Satyarthee GD, Moscote-Salazar LR. Extra-axial giant falcine ependymoma with ultra-rapid growth in child: uncommon entity with literature review. J Pediatr Neurosci 2016;11:324.

[21] Karthigeyan M, Singhal P, Salunke P, *et al*. A missed differential in an extra-axial lesion with calvarial involvement. Ann Neurosci 2017;24: 207–11.

[22] Nagayasu MA, Fukushima T, Matsumoto F, *et al*. Supratentorial extra-axial RELA fusion-positive ependymoma misdiagnosed as meningioma by intraoperative histological and cytological examinations: a case report. J Med Case Rep 2022;16:312.

[23] Yafang D, Zonghui L. Cerebellopontine angle anaplastic ependymoma: a case report and literature. Cancer 2017;5:17–24.

[24] 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.

[25] Mansur DB, Perry A, Rajaram V, *et al*. Postoperative radiation therapy for grade II and III intracranial ependymoma. Int J Radiation Oncol Biol Physics 2005;61:387–91.