Clear cell ependymoma with an atypical localisation mimicking intracranial glial tumour: A case report

Okan Turk[*], **Burak Tahmazoglu**[**], **Can Yaldiz**[***] **and Veysel Antar**[*]

[*]Department of Neurosurgery, Istanbul Training and Research Hospital, Istanbul, Turkey., [**]Department of Neurosurgery, Cerrahpasa Medical Faculty , Istanbul University, Istanbul, Turkey., [***]Department of Neurosurgery, Sakarya Training and Research Hospital, Sakarya, Turkey.

ABSTRACT Introduction: Ependymomas are tumours of the central nervous system, originating from the ependymal cells which line the ventricles and the central canal of the spinal cord. Less than one-third of all ependymomas are located supratentorially, most of the time near the ventricle walls around the trigone area. **Case:** A 34-year-old patient came into the outpatient clinic with a headache and seizure. After radiological examination, she was operated on for an extra-axial lesion located next to the left frontal gyrus. Pathological examination revealed a clear-cell ependymoma, WHO grade II, after which follow-up without any additional therapy was decided since the patient was young and total resection had been achieved. **Conclusion:** Because of low incidence, radiological diagnosis is difficult. Even if total resection has been achieved, long term follow-up should be planned because of high rates of recurrence.

KEYWORDS Intracranial, Clear-cell, ependymoma

Introduction

Ependymomas are tumours of the central nervous system, originating from the ependymal cells which line the ventricles and the central canal of the spinal cord. Although they are more frequently seen in the paediatric population, they may be found in adults as well [1]. Even though ependymomas are usually located infratentorially, in adults, they tend to have supratentorial localisations. Slow-growing lesions that seldom show signs of anaplasia, ependymomas have an 80% recurrence rate after treatment [2]. Most frequent localisation for intracranial ependymomas is the fourth ventricle [3]. Less than one-third of all ependymomas are located supratentorially, most of the time near the ventricle walls around the trigone area [1,2]. Extra-axial localisation for a supratentorial ependymoma is extremely rare [4]. In this paper, we found it noteworthy that an extra axially located supratentorial ependymoma mimicked the magnetic resonance imaging (MRI) findings of a neuroglial tumour.

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[1]Department of Neurosurgery, Istanbul Training and Research Hospital, Istanbul, Turkey.; Email:drokanturk@yahoo.com

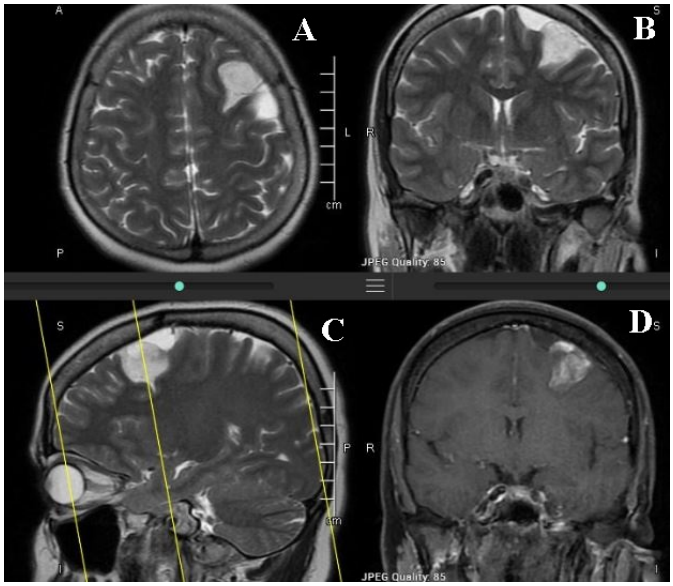

Figure 1: Pre-operative MRI showing the tumour.

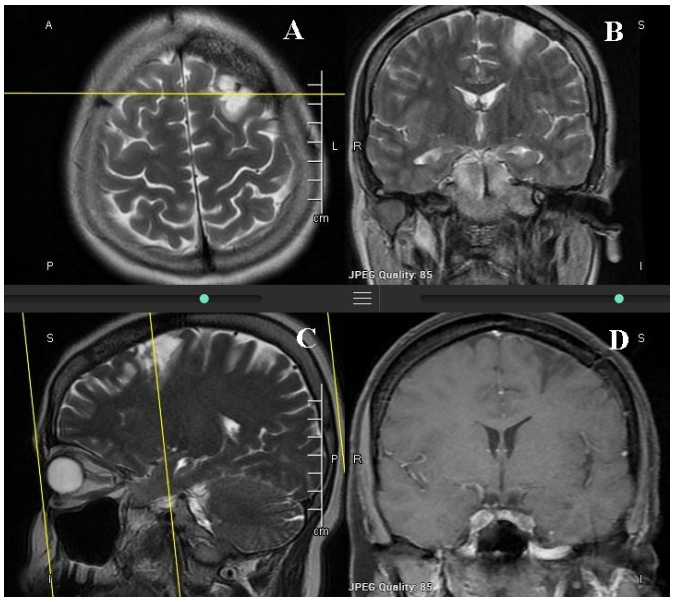

Figure 2: Postoperative MRI showed a total resection.

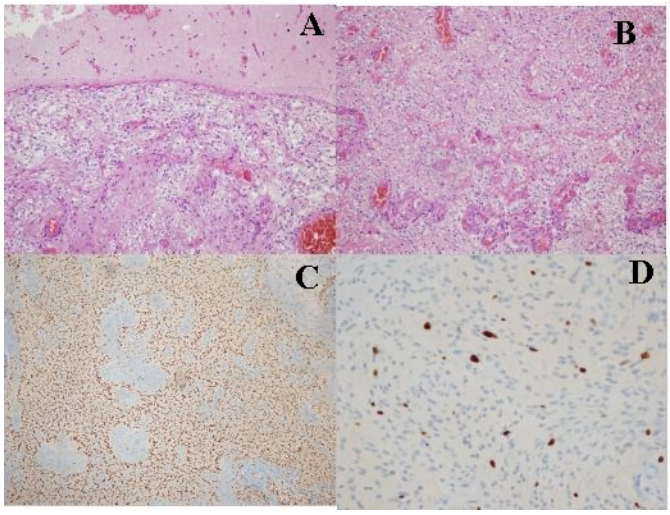

Figure 3: A) HEX200 tumoral infiltration with expansive growth, B) HEX200 Tumor infiltration-vascular endothelial proliferation consisting of clear cells, C) OLIG2X200 immunohistochemical OLIG2, D) KI67X499.

Case Report

A 34-year-old female patient came into the outpatient clinic with complaints of a headache and seizure. She had a history of a mild headache for the past year that in the last two months progressed. After a seizure episode, a cranial imaging study was performed, after which she was found to have an intracranial lesion. She was found to have no neurological deficits upon physical examination.

Medical history was non-significant, and there were no pathological findings in her haematological workup. Cranial MRI showed a 54x36x12mm heterogenous extra-axial lesion next to the left superior frontal gyrus that was pressing on the brain parenchyma, with a 22x20mm solid component that was hyperintense in T2W and T1W series and showed contrast enhancement (figure 1). The patient was operated on with an early diagnosis of the intraparenchymal neuroglial tumour. Postoperative MRI showed total resection was achieved (figure 2). On postoperative follow-up examination, the patient was found to have no neuro deficit.

Upon microscopic examination of the pathological studies, the tumour was made up of glial cells with clear cytoplasm and had an expansive growth pattern. Every field showed lobular cell groups made up of fibrovascular compartments rich with vascular structures. The cells of a tumour generally had round cells, vesicular chromatin structure and clear cytoplasms. In a couple of HPF, pseudorosette formation was seen. Moderate levels of pleomorphism were seen. Focal vascular endothelial proliferation was seen, but necrosis was not found. Immunohistochemical findings showed tumour cell nuclei were positive for olig-2 and sparsely positive for EMA. Moreover, IDH-1: negative, Ki67 index: % 2-3 specimen was pathologically diagnosed as clear-cell ependymoma, WHO grade II (figure 3).

Follow-up without any additional therapy was decided since the patient was young and total resection had been achieved.

Discussion

Ependymomas are tumours of the central nervous system, originating from the ependymal cells which line the ventricles and the central canal of the spinal cord. Usually, in paediatric cases, the tumour location is intracranial; in adults, on the other hand, the location is the medulla spinalis. When they are located intracranially, they are most likely to be situated in the infratentorial space, but especially in adults, they are found supratentorial [3]. As is our case, extra-axial ependymomas are extremely rare [3,4]. Furthermore, a clear-cell subtype of the ependymomas are rarer compared to other subtypes, and these subtypes are usually listed as WHO grade III ependymomas [5]. No mechanism for the development of these neoplasms has been set forth until now [3].

Other than the clinical differences between supratentorial, infratentorial and spinal ependymomas, there are also critical radiological differences [3]. Supratentorial ependymomas may cause headaches, seizures and focal neurological deficits [3] — the case in this paper presented with a seizure. In current practice, MRI is accepted as the gold standard in diagnosis and follow-up of intracranial lesions. Ependymomas are iso-hypointense in T1W series and hyperintense in T2W sequence. Their contrast enhancement characteristics are heterogeneous [1]. The MRI findings of the case in this paper were by these findings.

Total surgical resection is the preferred modal of treatment for supratentorial ependymomas. WHO has specified the role of radiotherapy in the treatment of ependymomas in previous studies [3]. Prognosis is related to tumour grade and resection limits. In our case, total surgical resection was achieved, and no further oncological treatment was planned. On 29 month follow-up, no recurrence or new seizure episodes were recorded.

Mansur et al. [6] argued in their study that surgical resection limit was the most important prognostic factor. They and the general literature on ependymomas proposed conventional radiotherapy because total resection could rarely be achieved. Oya et al. [7], Anh et al. [3] reported that whole spine radiotherapy for localised supratentorial ependymomas regardless of tumour grade would not be beneficial when adverse effects related to radiotherapy were considered. The prevailing opinion is that total surgical resection is the best treatment modality [3]. In case of a residual tumour or recurrence, adjuvant radiotherapy may be considered. There is no definitive evidence of the effective-

ness of chemotherapy [3]. In our case, because the patient was young and total surgical resection was achieved and considering the histopathological findings, we decided on close follow-up without any further treatment. After 29 months of follow-up, no recurrence was recorded.

Conclusion

Ependymomas of the convexities are extremely rare. Because of low incidence, they are difficult to diagnose using only radiological findings. Even if total surgical resection has been achieved, long term close follow-up must be planned because of the high probability of recurrence.

Competing Interests and Funding

All co-authors have seen and agree with the contents of the manuscript and there is no financial interest to report. We certify that the submission is not under review at any other publication.

Ethics committee approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee (name of institute/committee) and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

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