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Letter to the Editor

Cerebellar cystic glioblastomas: An uncommon presentation of a rare disease and clinical review

Dear Editor,

Cerebellar glioblastomas (GBM) are uncommon posterior fossa lesions found in adults. Despite intratumoral apoplectic events being common neurosurgical occurrences, it has rarely been described in posterior fossa GBM's. The impact of intratumoral cysts has been a matter of discussion, and their presence in the posterior fossa has been related to benign lesions such as pilocytic astrocytoma's, haemangioblastoma's, or malignant lesions like metastasis, very rarely, malignant primary glial lesions. An indolent clinical manifestation is not typical of GBM's, particularly when considering a posterior fossa cystic lesion. Given the previous assumptions, we present a case of an intratumoral bleeding into a cystic, well-defined lesion diagnosed as a primary GBM.

A 64-year-old female patient was referred to the neurosurgical outpatient clinic with a 2-week history of dysarthria and dysphagia. She had no other neurological deficit. A computer tomography (CT) scan revealed a hypodense lesion in the posterior fossa with minimal mass effect. The follow on magnetic resonance image (MRI) scan showed a cystic lesion with no surrounding oedema, initially interpreted as benign lesion (Fig. 1A–D). No treatment was addressed at this time and the patient was followed up in clinic.

Two months later, she presented to the Emergency Department with a decreased consciousness state after suffering from a severe headache. Imaging revealed intra-lesional haemorrhage and acute hydrocephalus. An external ventricular drainage (EVD), posterior fossa decompressive craniectomy and later surgical evacuation of the tumour was undertaken. Post operatively, her conscious state, de novo parinaud syndrome, dysarthria and dysphagia improved. She was discharged home with a modified Rankin score of 2 and residual ataxia. The postoperative CT scan revealed complete removal of the haemorrhagic component.

The pathological specimen removed for histological examination was extensively haemorrhagic (Fig. 1E–H) and the diagnosis of GBM, IDH- wildtype was confirmed. She was started on adjuvant treatment according to the Stupp protocol.

The origin of cerebellar cystic GBM is not completely understood. Given the proportion of neurons within the cerebellum in comparison to the number of neurons in the supratentorial compartment, around 10% of GBM's [1] should be located in this region, a number far superior to the 0.4–3.4% that has been cited [2]. Some of the literature has described cerebellar astrocytes having a lesser tendency for malignant transformation, even though this is an assumption based on the above epidemiology with no clear explanation behind it [3].

The implication of the cystic morphology in a patient's outcome has been a matter of debate. Different molecules have been identified, namely; lactate, glutamate and phosphate. It is speculated that these cysts may serve as a reservoir of nutrients supporting tumour growth, or be an accumulation of tumour secretion products [4]. Two main studies have compared the outcomes in patients with cystic versus non-cystic gliomas. Maldaun et al. [5], performed a tumour-volume matched cohort study and identified no differences in overall survival, but a significant difference in favour of cystic GBMs for time to recurrence (mean - 7.6 months, 95% CI [0.01–18 months] versus non-cystic GBMs - 4.2 months, 95% CI [1.8–6.6 months], Log-Rank test $p = .04$). Kaur et al. [6], also did not find any difference in survival between both groups.

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The posterior fossa GBM has a different identity from its supratentorial counterpart. They tend to be smaller and multifocal at diagnosis, H3K27M or NF1 related and lack EGFR amplification. The patients are often younger, the disease usually progresses with leptomeningeal or supratentorial dissemination and the overall survival is shorter [1]. When considering the radiological appearances, Occhiogrosso et al. [7], found little peritumoral oedema in patients with cerebellar GBM, and Zito et al. [8], stated this has diagnostic value in differentiating GBM from metastasis of the cerebellum via peritumoral edema or mass effect.

Intratumoural bleeding in GBM's is commonly seen with the proliferation through reticular capillaries and the lack of supportive stroma which was first described by Kondziolka et al. [9], (19.3% of patients had intratumoral bleeding). Anticoagulation and hypertension don't seem to be important risk factors when there is an underlying malignancy [8]. Considering the most common causes of posterior fossa hematomas are hypertension and anticoagulation, and the most common causes of posterior fossa tumours are metastasis and hemangioblastoma, the present case is unexpected, but should be consider in the differential diagnosis.

Haemorrhagic cystic posterior fossa glioblastoma's are a rare

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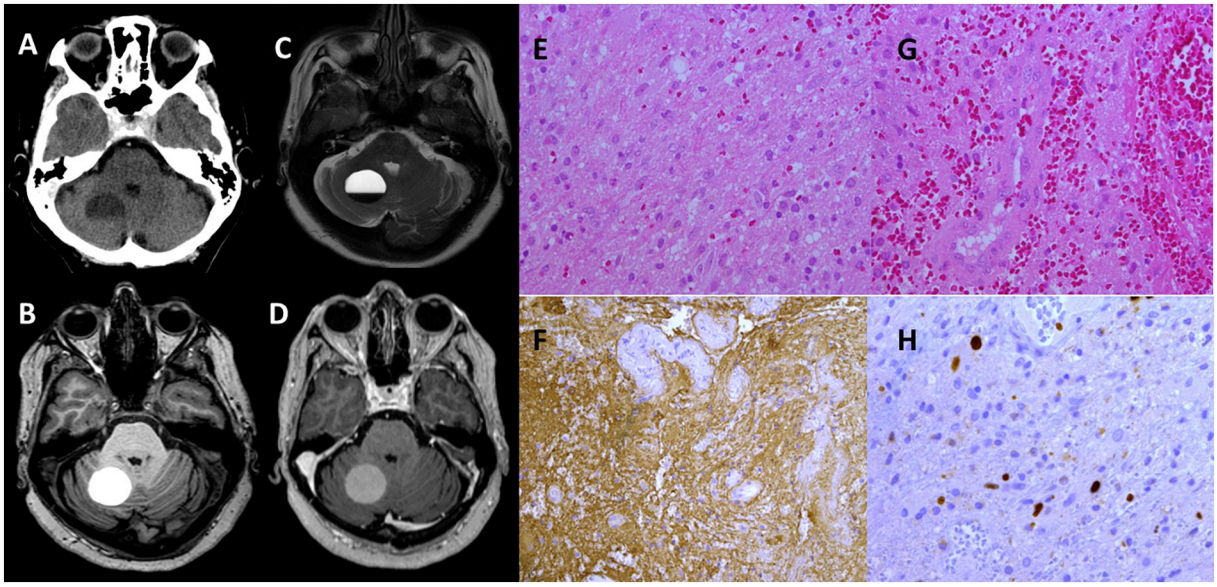

Fig. 1. A - Non-contrast axial CT Scan revealing a mixed density lesion with a probable cystic component and a fluid level; B - non-contrast T1-weighted; C - T2-weighted; D - T1-weighted post-GAD; E - Hypercellular glial tissue composed of fibrillary and sporadic pilocytic astrocytes with hyperchromatic and pleomorphic nuclei, mitotic features and microvascular proliferation (HE ×40); F - GFAP immunoreactivity, ATRX (no mutation) and P53 immunoreactivity but IDH1 immunonegativity; G - Endothelial proliferation of the tumour vascularization (HE ×40); H - High proliferative index of 20% (Ki67 ×40). Further sequencing showed no IDH1, IDH2, BRAF, H3F3A, hTERT mutations and unmethylated MGMT.

differential. Nevertheless, an aggressive diagnostic attitude should be supported towards unclear posterior fossa lesions.

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José Pedro Lavrador[a], José Hipólito Reis[b], Diogo Belo[b], Shami Acharya[a,*], Cláudia C. Faria[b], José Pimentel[c], Manuel Herculano Carvalho[b]
[a] *Department of Neurosurgery, King's College Hospital Foundation Trust, London, UK*
[b] *Department of Neurosurgery, Hospital Santa Maria, Centro Hospitalar Lisboa Norte, Lisboa, Portugal*
[c] *Laboratory of Neuropathology, Neurology Department, Centro Hospitalar Lisboa Norte, Lisboa, Portugal*
E-mail address: acharya.sy@gmail.com (S. Acharya).

[*] Corresponding author at: 39 Gover Court, Paradise Road, London SW46QN, United Kingdom.