

Dorsally exophytic glioblastoma arising from the medulla oblongata in an adult presenting as 4th ventricular mass

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ABSTRACT

Brainstem gliomas are relatively rare in adults (<2% of all gliomas). Exophytic gliomas are focal brainstem lesions, which project into the 4th ventricle or cerebellopontine angles. These exophytic lesions are usually of low-grade histology (pilocytic astrocytoma or ganglioglioma) and have a relatively better outcome compared with brainstem gliomas as a whole. Glioblastoma is the commonest primary glial cell neoplasm and mostly occurs in the supratentorial compartment. It is rather uncommon in the brainstem and seldom has been described as having an exophytic growth pattern. Here we describe an exophytic brainstem glioblastoma arising from the medulla oblongata in a 55-year-old lady who presented with a 4th ventricular mass, and present a brief review of the literature. Till now, six cases of glioblastoma arising from the medulla oblongata have been reported. So, ours is the seventh such report. To the best of our knowledge, it also happens to be the sixth reported case of dorsally exophytic brainstem glioblastoma till date.

Key words: Adult, brainstem glioblastoma, dorsally exophytic, surgery, 4th ventricular mass

Introduction

Brainstem gliomas are common in the paediatric population, constituting around 10%-20% of all gliomas in this age group.^[1] They represent a formidable surgical challenge by virtue of their deep location and presence of vital neural centres within. This partly explains the poor prognosis associated with these tumours.

In contrast to children, brainstem glioma is a rarity in adulthood (<2% of all gliomas). Studies indicate that adult brainstem gliomas tend to have a better survival than children

in spite of the fact that they more often harbour high-grade lesions.^[2]

Dorsally exophytic brainstem gliomas are focal brainstem lesions, which are usually low-grade tumours on histology and are amenable to surgical excision, with a comparatively better survival than the rest.^[3] Much more common in the supratentorial location, glioblastoma arising from the medulla oblongata is an extremely rare occurrence.^[4] To the best of our knowledge, only six cases of glioblastoma affecting the medulla have been described in the literature till now.^[4-8]

Here we describe the case of a 55-year-old lady with an exophytic brainstem glioma arising from the lower part of the 4th ventricular floor (medulla), and discuss our findings in the light of relevant literature.

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Case Report

History and presentation

A 55-year-old lady presented to us with increasing bifrontal headache and recurrent vomiting of five-month duration. She did not have any difficulty in walking or swallowing. There was no history of seizure, change in voice or any limb weakness. On examination, she had bilateral papilloedema without any other neurological deficits.

Computed tomography of the brain revealed an ill-defined hypodense mass in the midline posterior fossa with mild heterogeneous contrast enhancement. There was mild hydrocephalus and lack of intra-tumoral calcification [Figure 1a].

Gadolinium-enhanced magnetic resonance imaging of the brain revealed a heterogeneously enhancing irregular

intra 4th ventricular mass with a poor plane of cleavage from the surrounding structures. Ventriculomegaly with periventricular lucency (PVL) was clear in T2 weighted images. The mass was seen extending caudally up to the foramen magnum [Figure 1b-d].

Clinico-radiological picture thus strongly indicated the diagnosis of ependymoma.

Operation and post-operative course

The tumour was approached by midline sub-occipital craniectomy. The dura appeared tense. Following Y-shaped durotomy, the cerebrospinal fluid (CSF) was released from the cistern magna to relax the brain. The vermis was lifted upwards after partially dividing its lower pole. The tumour was seen as a dirt grey, friable mass with broad-based adherence to the medulla part of the floor of the 4th ventricle. The tumour

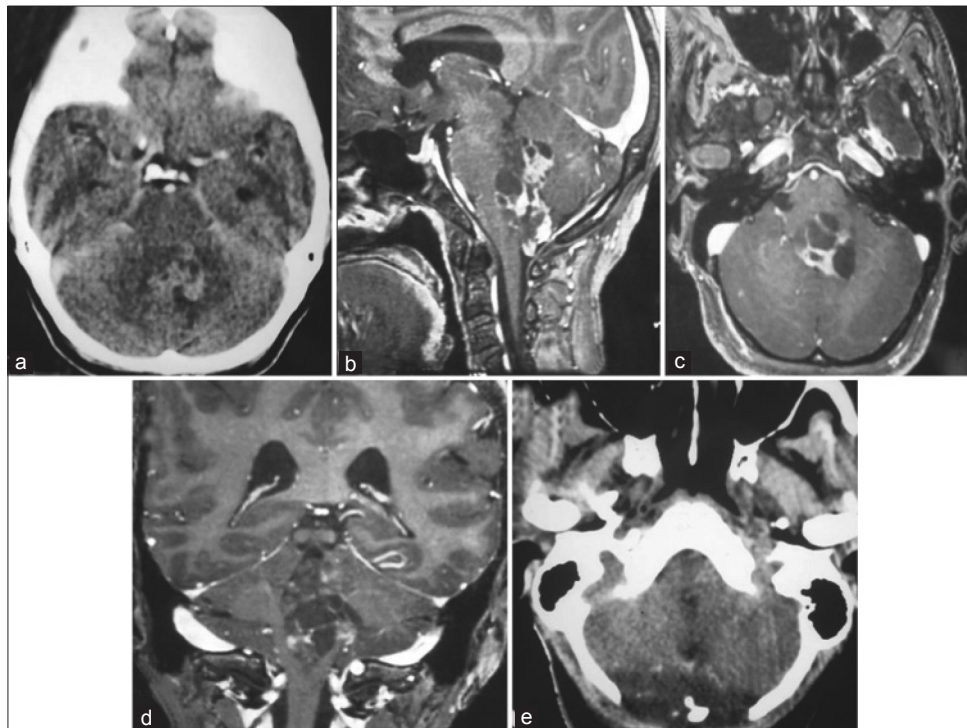


Figure 1: (a) A contrast-enhanced CT head shows an irregular, heterogeneously enhancing 4th ventricular mass without any plane of cleavage all around. (b) A contrast-enhanced magnetic resonance imaging head in the sagittal plane shows the heterogeneous, dorsally exophytic, irregularly enhancing mass within the 4th ventricle without any clear plane of cleavage from the brainstem. Inferiorly, it is seen extending through the foramen of Magendie. (c) Axial and (d) coronal contrast MR images showing the exophytic mass in the midline without any CP angle extension. (e) A post-operative CT head showing the craniectomy defect without any contrast-enhancing mass in the region of the brainstem

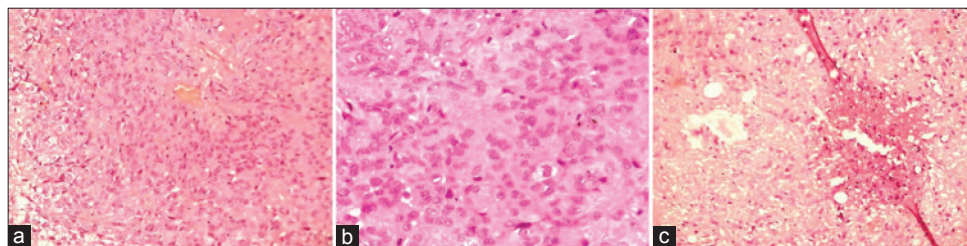


Figure 2: (a) Haematoxylin and eosin staining showing a tumour disposed in sheets of moderately pleomorphic cells on a fibrillary background (x200). High cellularity increased (b) mitosis (x400) and (c) necrosis (x200)

was removed except the part adherent to the brainstem, which was deliberately left behind. The CSF pathway was established and haemostasis was achieved.

The patient recovered uneventfully following surgery except mild paresis of bilateral lateral gaze, which recovered gradually. Post-operative scan showed no residual enhancement [Figure 1e]. Histopathological features were consistent with diagnosis of conventional glioblastoma [Figure 2a-c]. At discharge we advised her radiotherapy.

At 3-month follow-up she had completed her radiotherapy (a total of 60 Gy over 30 fractions) and was doing well. She was advised chemotherapy (temozolomide) but relatives could not afford the same. So, she was advised regular follow-up.

Discussion

Epidemiology

Brainstem gliomas are common in children where they account for nearly 10-20% of all gliomas, but it is a rarity in adults (constituting only less than 2% of all gliomas).^[1,2] These tumours represent a formidable surgical challenge because of their deep location within the brain along with presence of numerous nerve tracts, cranial nerve nuclei and various vital centres. Pathologically, these tumours are heterogeneous. Whereas most childhood brainstem gliomas tend to be benign, malignant histology predominates in adults.^[2]

Brainstem gliomas have been variously classified in the literature. Epstein and Farmer^[9] have classified them into diffuse, focal, exophytic and cervico-medullary types. Guillamo *et al.*^[2] analyzing a large number of adult brainstem gliomas, have classified them into adult diffuse infiltrative low-grade glioma, adult malignant brainstem glioma, focal tectal brainstem glioma and others. Exophytic brainstem gliomas (Epstein classification) are focal lesions arising from the sub-ependymal glial tissues of the 4th ventricular floor, with subsequent transependymal projection into the ventricular cavity or the cerebellopontine angles. It is believed that growth of brainstem tumours is guided by the structures within the brainstem, leading to an exophytic growth pattern.^[9] These lesions are distinct from the other types of brainstem gliomas

in many aspects. These are usually low-grade lesions on histology (usually pilocytic astrocytoma or ganglioglioma), often show brilliant post-contrast enhancement, are amenable to surgical excision and most importantly have a comparatively better outcome.^[3]

Brainstem glioblastomas are very rare when compared with supratentorial glioblastomas. In one of the largest series on adult brainstem gliomas, Guillamo *et al.*^[2] reported only four cases of glioblastoma (12.5%). It is further unusual to have a brainstem glioblastoma with a dorsally exophytic growth pattern. None of the patients described by Guillamo *et al.*^[2] and Pollack *et al.*^[3] had a dorsally exophytic brainstem glioblastoma. Considering the fact that dorsally exophytic brainstem gliomas almost always harbour a low-grade histology and as high as 27% of low-grade brainstem gliomas are estimated to progress to higher grades,^[2] there are reasons to believe that exophytic brainstem glioblastomas could actually be secondary glioblastomas.

An extensive literature review showed six cases of brainstem glioblastoma arising from the medulla oblongata till date of which four were adults. We also identified five cases of exophytic brainstem glioblastoma. The table summarises these cases [Table 1].

Clinico-radiological profile

Clinically, brainstem glioblastomas present with a short duration of symptoms, which mainly include multiple cranial nerve palsies and long tract signs. However, our patient presented with symptoms of obstructive hydrocephalus only. The symptomatology was also in contrast to exophytic brainstem gliomas on the whole where usual presentation is gait disturbance (due to either corticospinal or cerebellar involvement). This is followed by headache, nausea and vomiting, and lower cranial nerve dysfunction. Visual disturbance, vomiting and headache are more frequent in children. By contrast, gait disturbance, headache, vertigo and lower cranial nerve dysfunction are more common in adults. Atypical presentations reported include symptoms of a cerebellopontine angle mass, myasthenia gravis and intractable hiccups.^[11]

Table 1: The Summary of the exophytic brainstem glioblastomas reported till date

Author (year)	Age (years)/sex	Histology	Exophytic extension into	Follow-up and status	Origin
Salunke <i>et al.</i> ^[4]	59/M	Conventional glioblastoma	CP angle	-	Medulla
Chotai <i>et al.</i> ^[4]	51/F	Conventional glioblastoma	4 th ventricle	19 months; disease progressed	Medulla
Queiroz <i>et al.</i> ^[5]	13/-	Did not specify	-	Died	Medulla
Abbott <i>et al.</i> ^[6]	Paediatric/-	Did not specify	-	Died after 3 months of surgery	Medulla
Kyoshima <i>et al.</i> ^[7]	55/-	Conventional glioblastoma	4 th ventricle	2 years 3 months	Medulla
Luetjens <i>et al.</i> ^[8]	40/M	Giant cell glioblastoma	4 th ventricle	2 years; no recurrence	Medulla
Rasalingam <i>et al.</i> ^[10]	9/M	Conventional glioblastoma	CP angle	Death after 2 months of surgery	Pons
Present case	55/F	Conventional glioblastoma	4 th ventricle	3 months; clinically stable	Medulla

From the radiological point of view, glioblastomas in the brainstem show necrosis and irregular enhancement like any other parts of the brain^[2]. In our patient, an in-homogeneously enhancing, ill-defined intra 4th ventricular mass extending caudally through the foramen of Magendie was seen. This closely resembled the picture of ependymoma (history also suggested the same). In addition to ependymoma, sub-ependymoma, metastasis and pleomorphic xanthoastrocytomas are the usual radiological differentials.^[8] Hence, imaging may be confusing at times.

Treatment and prognosis

We deliberately performed a sub-total tumour resection (removed the exophytic part and left the part densely adherent to the medulla) in our patient and subjected her to post-operative External Beam Radiation Therapy (EBRT). The patient had financial constraints, so temozolomide could not be administered. Although the standard treatment of glioblastoma includes maximal surgical resection followed by concurrent chemo-radiotherapy (with temozolomide),^[12] a conservative resection is very much justified in an area like the medulla oblongata. This not only avoids brainstem dysfunction, but prevents impairment of quality of life as well. Moreover, any left over tumour can be managed with adjuvant therapy.^[13] Kyoshima *et al.*^[7] have, however, advocated aggressive resection of glioblastoma of the medulla oblongata for better outcome. Analyzing the surgical anatomy, they have stated that laterally located tumours within the medulla can be removed totally with relative impunity, but midline tumours should be dealt with caution, as there are high chances of post-operative morbidity.

As far as the prognosis is concerned, Guillamo *et al.* have stated that a non-malignant brainstem glioma has a better prognosis in adults, but the same cannot be said for their malignant counterparts (median survival about 11.2 months).^[2] Although earlier reports showed a dismal outcome in these patients, in three recent papers, overall survival in the tune of nearly 2 years has been reported.^[1,7,8] Based on our case and review of other cases, we think that one should try to perform maximum possible surgical debulking of these tumours located in such vital areas followed by adjuvant radio- and chemotherapy for a better outcome. If it has diffusely infiltrated into the brain stem, however, a conservative approach rather than radical surgical excision brings about a better functional outcome.

Conclusion

Exophytic brainstem glioblastoma in adult is very rare, but may pose diagnostic difficulty like in our case. Hence, it should be considered as a differential diagnosis while evaluating 4th ventricular lesions in adults. The treatment is like glioblastoma elsewhere in the brain. Their prognosis is not exactly known, but appears to be slightly better than glioblastoma on the whole.

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Conflicts of interest

There are no conflicts of interest.

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