

Malignant cerebellar peduncle lesions - rapid progression and poor outcome

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Abstract

Background: Tumors arising from cerebellar peduncle are extremely rare and behave aggressively. The inclusion of these into either cerebellar or brainstem gliomas is contentious.

Case Description: We performed clinicopathological review of three patients treated at our institute and surveyed the literature for previous such reported cases. Mean duration of symptoms in our patients was 2 weeks. Subtotal tumor resection was performed in two patients while the third underwent stereotactic biopsy followed by chemoradiotherapy. Histopathology revealed glioblastoma in initial two patients and medulloblastoma Grade IV in the third. The two patients who underwent surgical excision succumbed to the illness within 2 days and a month, respectively.

Conclusion: Malignant cerebellar peduncular lesions have poor overall survival despite surgical debulking. It is not confirmed whether these tumors should be considered as cerebellar lesions or brainstem gliomas due to aggressive clinical behavior, and so the ideal line of management is not yet known.

Key Words: Glioblastoma cerebellar peduncle, malignant peduncular lesions, outcome cerebellar lesions

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INTRODUCTION

Primary cerebellar peduncle lesions are defined as the ones that arise directly from the peduncle and spread to involve the neighboring cerebellum and brainstem vital areas. Malignant lesions at this location are rare. It is a dilemma whether they behave as cerebellar tumors or as brainstem tumors. This ignites controversy of whether radical surgery is beneficial or histological diagnosis followed by radiotherapy should be attempted. There is scarce literature on the same and prognosis remains dismal despite surgery and radiotherapy. These tumors are known for their low overall survival (OS) and early

recurrence.^[7] We report a series of three such patients and discuss the clinicopathological features of the same.

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CASE REPORTS

Case 1

A 10-year-old male presented with complaints of gait disturbance and vomiting for 2 weeks. On examination, he was drowsy and had left sided cerebellar signs and a weak gag reflex on left side. Magnetic resonance imaging (MRI) [Figure 1] revealed solid-cystic lesion in left anterior cerebellum and cerebellar peduncle that was hypointense on T1-weighted (T1W), hyperintense on T2-weighted (T2W), had heterogenous contrast enhancement with surrounding edema. Midline suboccipital craniotomy was performed for tumor excision. Histology was suggestive of low-grade glioma. Thereafter, patient was lost to follow-up and presented 3 months later in unconscious state with a large recurrence and hydrocephalus. A critical review of previous biopsy showed few elements of atypical cells with high mitotic activity. The patient underwent a ventriculoperitoneal shunt followed by reexploration. Intraoperatively the tumor was vascular, infiltrating surrounding cerebellum with areas of necrosis. Histopathology revealed glioblastoma (World Health Organization [WHO] Grade IV) with highly cellular tumor showing nuclear pleomorphism and endocapillary proliferation. The patient did not gain consciousness after surgery. Computed tomography (CT) was suggestive of hypodensity and edema in brainstem. Further neurological deterioration was noticed next day with episodes of tachycardia and apnea. The patient finally succumbed to illness due to brainstem edema after 2 days.

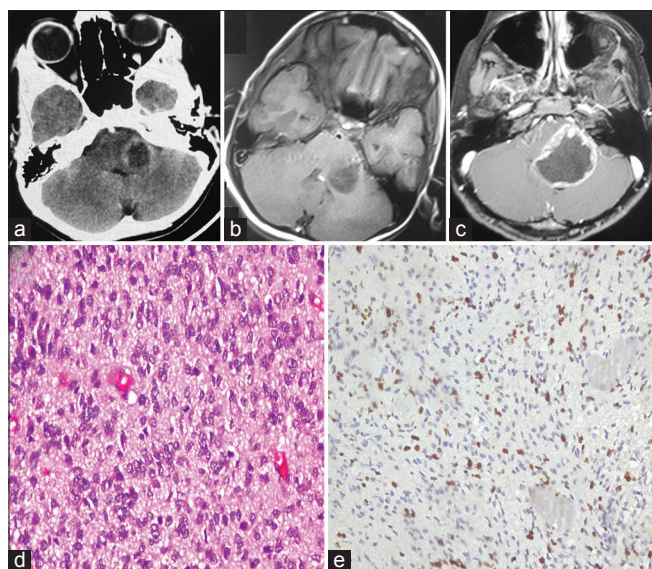


Figure 1: (a) Noncontrast computed tomography (axial) showing hypodense lesion at left cerebellar peduncle. (b) T1-weighted contrast axial image showing nonenhancing lesion before first surgery. (c) Axial T1-weighted contrast image at recurrence showing solid-cystic lesion at peduncle. (d) Photomicrograph showing tumor cells with nuclear pleomorphism, high mitotic activity (H and E, $\times 40$). (e) Ki-67 immunostain showing very high proliferation index (IP, $\times 40$)

Case 2

A 9-year-old female presented with complaint of diplopia on far vision and facial deviation for 1 week. Examination revealed gait ataxia, left gaze, facial paresis, and subtle left cerebellar signs. MRI [Figure 2] revealed a left peduncular lesion which was hypointense on T1W, hyperintense on T2W and had heterogenous enhancement on contrast. Two days later, she became unconscious, when a repeat CT showed intratumoral bleed again with mass effect. She underwent midline suboccipital craniotomy and had a friable, vascular tumor with necrosis. Histopathology revealed small cell glioblastoma (WHO Grade IV) with highly cellular tumor showing palisading necrosis, nuclear pleomorphism. Tumor cells stained positive for glial fibrillary acidic protein. Despite adequate decompression, the patient did not show clinical improvement. CT was suggestive of brainstem edema. There were scarce spontaneous respiratory efforts, pupil reaction was absent, and stimulus yielded minimal flexion response. She finally succumbed to illness after 1 month due to brainstem edema.

Case 3

A 30-year-old male patient presented with complaint of gait ataxia and visual blurring for 3 weeks. Examination revealed left cerebellar signs and a brun's nystagmus with coarse component to left. CT head demonstrated a hyperdense lesion in left anterior cerebellum spreading to middle cerebellar peduncle. MRI [Figure 3] demonstrated lesion that was hypointense on T1W, iso- to hyper-intense on T2W with necrotic foci at inferior end. Taking into consideration the close approximation to vital

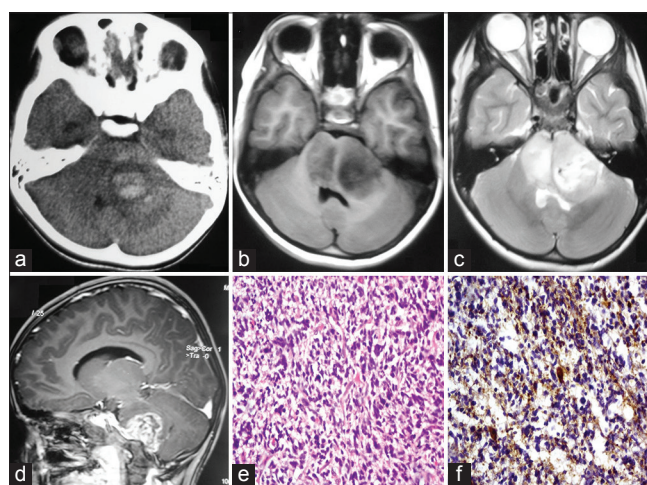


Figure 2: (a) Noncontrast computed tomography (axial) showing hemorrhagic lesion at left cerebellar peduncle. (b-d) Axial T1-weighted, axial T2-weighted, and sagittal T1-weighted contrast magnetic resonance imaging showing lesion hypointense on T1, heterogeneously hyperintense on T2 and dense contrast enhancing with edema and infiltrating pons. (e) Photomicrograph showing highly cellular glial tumor with high nuclear pleomorphism (H and E, $\times 40$). (f) Tumor cell shows glial fibrillary acidic protein positive cytoplasmic processes (IP, $\times 40$)

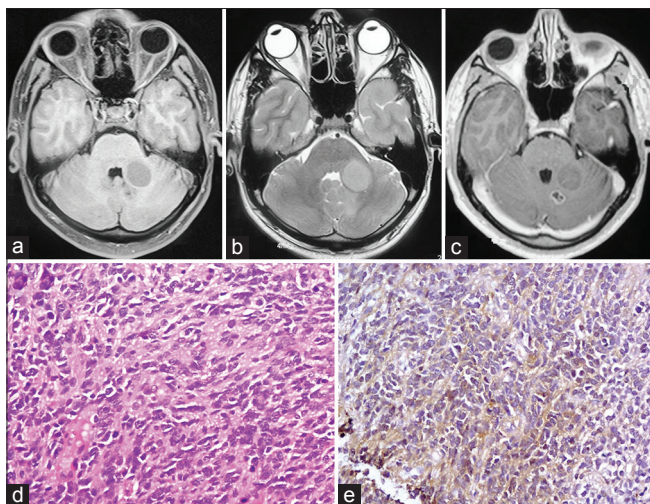


Figure 3: (a-c) T1-weighted, T2-weighted, and T1-weighted contrast axial images showing well-defined lesion at left middle cerebellar peduncle hypointense on T1, isointense on T2, predominantly nonenhancing on contrast with small posterior nodule showing ring enhancement and necrosis. (d) Photomicrograph showing cellular tumor composed of round, blue cells with high nuclear:cytoplasmic ratio and high mitotic activity and rosette formation (H and E, $\times 40$). (e) Tumor cells show faint cytoplasmic synaptophysin positivity (IP, $\times 40$)

brainstem area, he was taken up for stereotactic biopsy. Histopathology revealed medulloblastoma (Grade IV) with high mitotic activity, high nuclear:cytoplasmic ratio, and round cells. The tumor stained positive for synaptophysin and neuron-specific enolase. He received adjuvant radiotherapy and concurrent chemotherapy with cisplatin and etoposide. After ten weeks, he presented with headache and vomiting. Repeat MRI revealed slight increase in edema and hydrocephalus for which a ventriculoperitoneal shunt was performed. At follow-up of 6 months, he is conscious and independent for daily routine activities.

DISCUSSION

Glioblastomas in cerebellum account for $<5\%$ of all cerebellar astrocytomas.^[2,7,9] No apparent explanation accounts for the rarity of the infratentorial compartments as a site of origin for glioblastomas. Peduncular involvement in cerebellar lesions has been scarcely mentioned in the literature. The mean duration of symptoms in glioblastomas involving cerebellum is 4–5 weeks and OS of 9–15 months^[13] while it is only slightly better for the anaplastic gliomas.^[6,11,12] Symptomatology consists of headache, nausea and vomiting, diplopia, facial deviation, hearing disturbance, and gait ataxia. Brainstem signs are frequently present in these lesions.^[3] Compared to hemispheric tumors, the ones arising from cerebellar peduncle behave more aggressively, are invading in nature, and recur early.

Tumor frequently extends beyond the enhancing part seen on imaging. Dissemination is believed to occur along ventricular wall and subarachnoid space^[8-10,12] with reported rates of leptomeningeal spread ranging from 17% to 66%.^[7] While an individual can tolerate removal of an entire cerebellar hemisphere, its proximity to brainstem via the cerebellar peduncles makes it easy for tumor to spread to regions that would be considered unresectable. The poor outcome despite surgery and chemoradiotherapy in primary cerebellar lesions that recur at peduncle depicts the effect of involvement vital brainstem regions.^[4] The management of such aggressive lesions is a matter of debate as surgery may not provide a long-term survival.^[10] Considering the high leptomeningeal spread, role of craniospinal irradiation is well established.^[7] Peduncular involvement in medulloblastoma, as in case 3 in our study, has been reported earlier. The spread is believed to occur from flocculus that projects into cerebellopontine angle or residual of lateral medullary velum at this location.^[5] Migratory process of germinal cells in a lateral direction explains involvement of cerebellopontine angle in adult medulloblastoma.^[1] Although glioblastoma and medulloblastoma are entirely different pathologies, they may behave similarly as seen in our series where all lesions were high-grade (WHO Grade IV) with rapid dissemination.^[11]

The rapidity of onset of symptoms in these patients and previously reported patients harboring cerebellar high-grade lesions is alarming,^[10] with a mean duration of only 2 weeks. Two of three patients who underwent surgery died with a mean survival time of 2 weeks only. The third, after stereotactic biopsy, is under follow-up at 6 months. The overall scenario seems worse than that of supratentorial glioblastoma and brainstem gliomas, exact reason for which is not known. We believe that lesions which appear high grade and arise from cerebellar peduncle may have a natural history similar to aggressive brainstem gliomas.^[3] Extraneural spread similar to that seen in medulloblastoma may be present in cerebellar glioblastoma.^[11]

It is possible that the natural behavior and, therefore, the mode of failure of therapy, of cerebellar high-grade lesions invading cerebellar peduncle differ from hemispheric lesions. The truly aggressive nature of these lesions and the dismal survival in the presence of limited overall reported cases so far raises question of appropriate line of management.^[10] It is a controversy whether these lesions should be considered merely cerebellar pathologies or brainstem lesions with poor overall outcome.

Our experience with such high-grade lesions at cerebellar peduncle has made us believe that proper case selection

is imperative as aggressive surgical approach may be hazardous. With limited OS and associated risk of working in vital brainstem regions, biopsy followed by chemoradiotherapy may be a better option in few selected patients.

CONCLUSION

Malignant cerebellar peduncle lesions present with short history, progress rapidly, and may have a dismal outcome despite surgery. The small number of overall reported cases in literature leaves much to be sorted out in coming years. Although the number of patients in our study is small, certainly the focus of this controversy is the poorly defined natural history of high-grade lesions at cerebellar peduncle. We share our experience of managing such patients and together with a thorough literature review, put forth a rarely described entity, the ideal management of which is controversial.

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Conflicts of interest

There are no conflicts of interest.

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