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Case Report

Primary glioblastoma of cerebellopontine angle: The diagnostic dilemma of this rare case ☆

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ABSTRACT

Supratentorial glioblastoma is a common primary intracranial tumor in adults. High-grade glioma in cerebellopontine angle (CPA) is rare. We describe a case of adult-type diffuse high-grade glioma of CPA in a 49-year-old female that was managed surgically at our institute. Glioblastoma is an infiltrating type of glioma (WHO grade 4). MRI helped in characterizing the lesion; however, diagnosis was confirmed on histopathology. This report aims at the imaging pattern of primary adult-type diffuse high-grade glioma (WHO grade 4) at cerebellopontine angle.

Introduction

Most tumors in cerebellopontine angle are located extra-axially. These are usually acoustic schwannoma or meningiomas. Other less common are epidermoid tumors, metastases, and arachnoid cyst. Other literature also describes gliomas in CP angle. However, high-grade gliomas of CPA are extremely rare. A lesion arising from brain parenchyma may become exophytic and manifest in the CPA. Signs and symptoms are present due to compression of fifth, seventh, and eighth cranial nerves and lateral aspect of pons and cerebellum. It is very crucial for the neurosurgeon to make the differential diagnosis between theses lesions because their treatment modalities, prognosis, and outcome are different. Since clinical manifestations are similar in most of these tumors, hence making an appropriate diagnosis is important for the management of these tumors.

☆ Competing Interests: There is no interest to declare.

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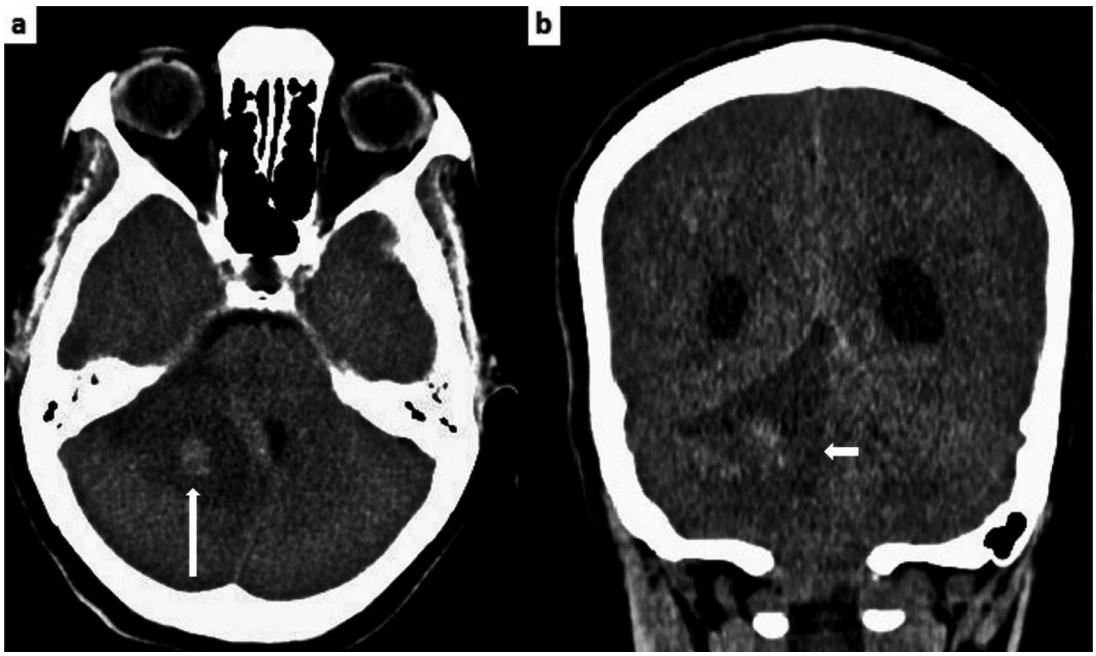

Fig. 1 – Noncontrast CT scan shows hypodense lesion in right cerebellopontine angle (CPA) with areas of hyperdensity, suggesting intratumoral bleed (long and short white arrows).

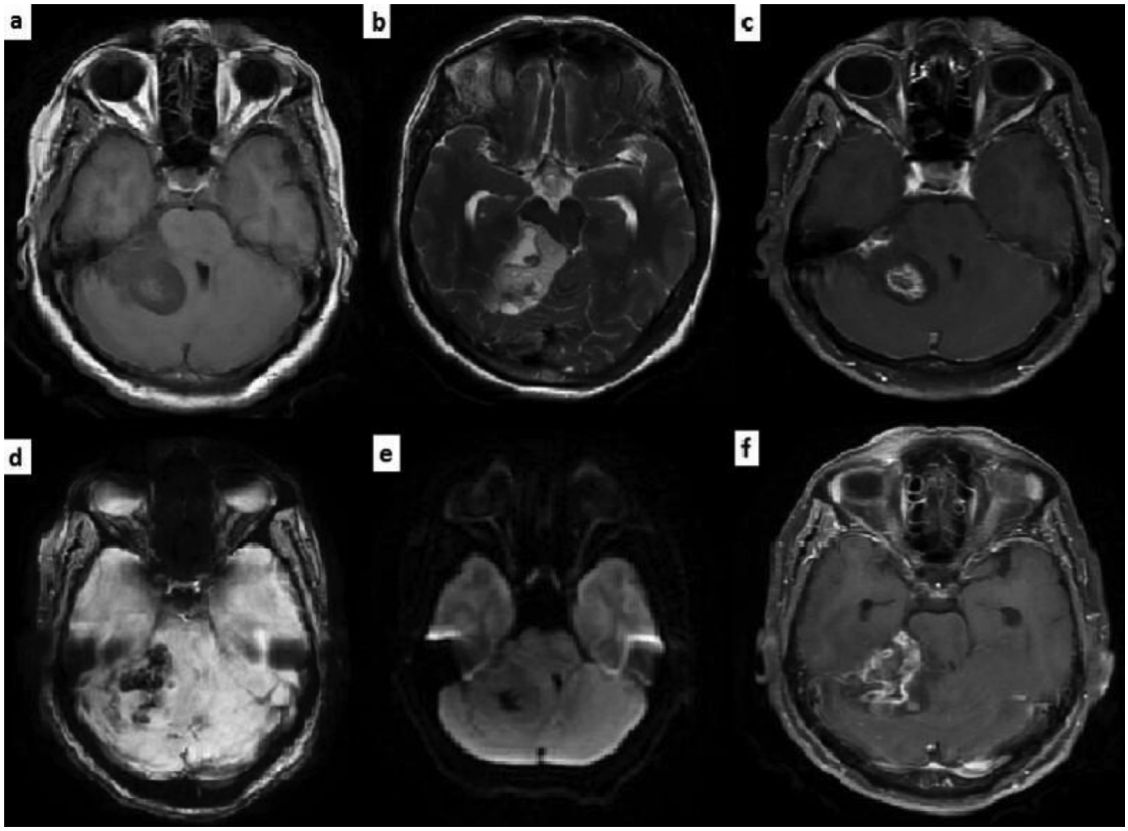

Fig. 2 – Brain magnetic resonance imaging axial images: (A) the tumor is heterogeneously hypointense on T1-weighted sequence with areas of hyperintensity, (B) heterogeneously hyperintense on T2-weighted sequence, (D) blooming on GRE sequence, (E) no restriction on diffusion-weighted sequence, (E,F) T1-weighted postgadolinium sequence showing heterogeneous contrast enhancement.

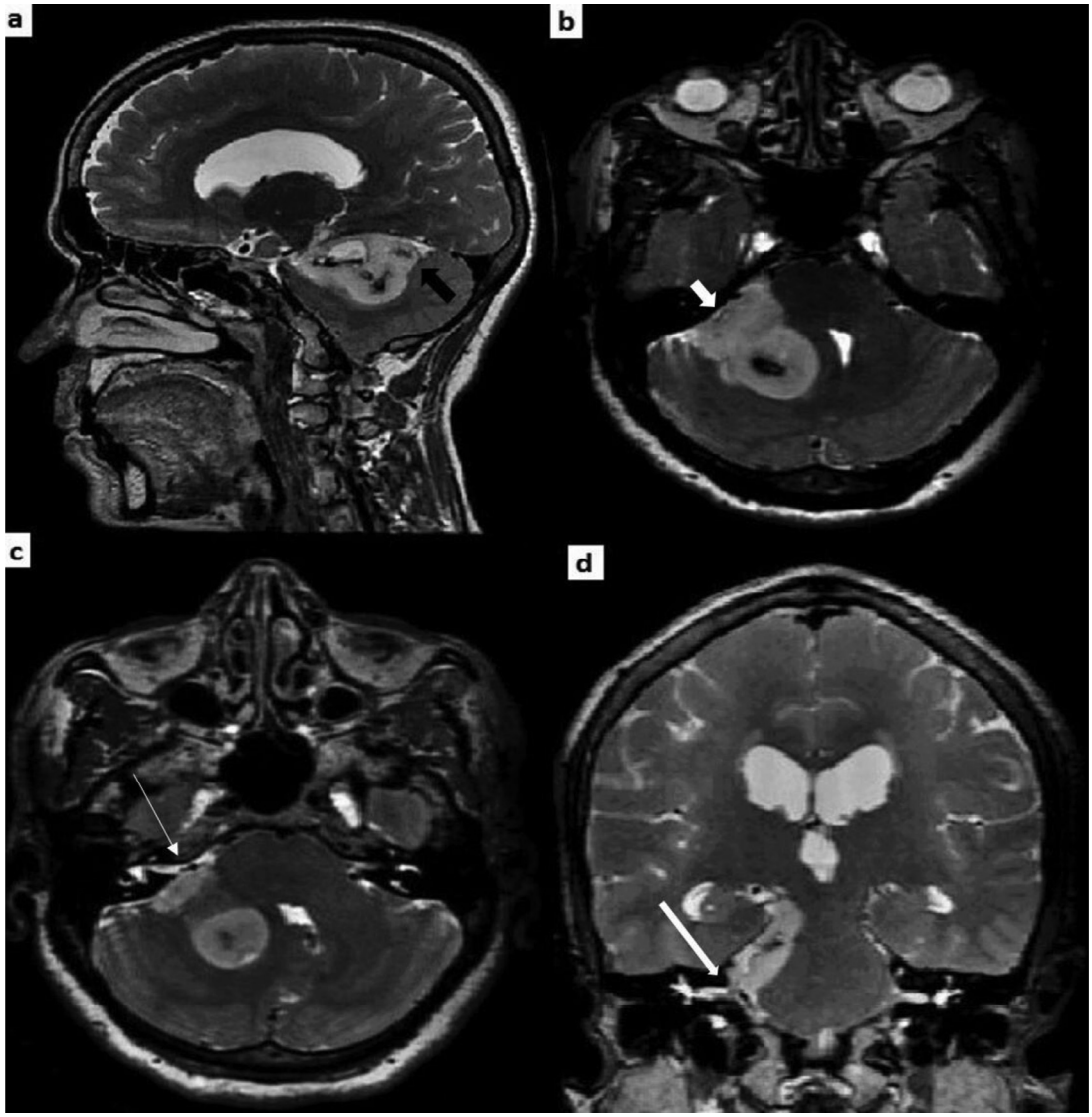

Fig. 3 – Brain magnetic resonance imaging sagittal, axial, and coronal T2-weighted sequence: (A, B) abutting the dura mater, mimicking a meningioma (short black and white arrows), (C, D) tumor in relation to seventh and eighth nerve complex (thin and thick long white arrows).

Case report

A 49-year-old lady presented with complaint of headache, giddiness, and difficulty in walking for 2 months. She had worsening vomiting and gait ataxia with swaying to right side for the last 10 days. On physical examination, she was afebrile with no acute distress. Her vitals were stable. On neurological examination, she was alert and fully oriented. Her pupils were equally reacting to light. Cranial examination revealed minimal seventh nerve paresis and reduced hearing on the right side. Finger to nose examination revealed dysmetria on right side. Sensory testing was grossly normal. A working diagnosis of lesion in CPA was made.

Brain computed tomography revealed heterogeneously hypodense lesion noted in right cerebellopontine angle, showing hyperdense areas within of blood attenuation suggesting intratumoral bleed (Hounsfield Unit- 56) (Fig. 1).

Magnetic resonance imaging demonstrated an ill-defined infratentorial lesion measuring ~ 45 × 47 × 31 mm in right CP angle with broad base on petrous bone. The lesion was infiltrating the cerebellum and was separated from the brain stem. The lesion appeared heterogeneously high signal intensity on T2-weighted MRI with areas of necrosis. It showed peripheral enhancement of the lesion on gadolinium enhanced T1-weighted image. Superimposed foci of blooming on GRE were noted suggesting intratumoral bleed with no restricted diffusion. Anteromedially, the lesion was compressing and displacing brainstem toward left side. It was effacing the fourth ventricle causing upstream hydrocephalus. Surrounding edema was also seen (Figs. 2 and 3). For differential diagnosis of the lesion, magnetic resonance spectroscopy (MRS) was taken, and it demonstrated increase in

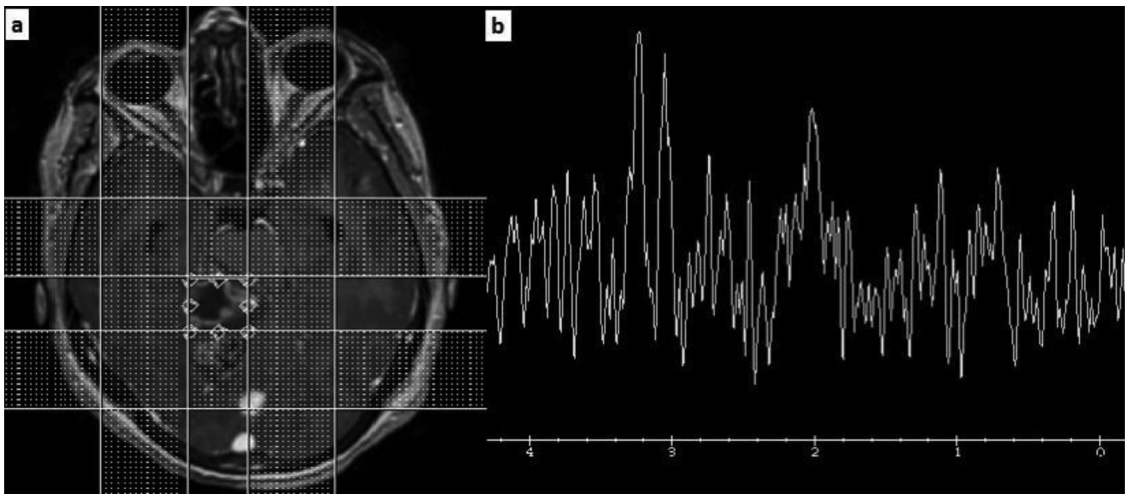

Fig. 4 – Magnetic resonance spectroscopy illustrates increase in choline and creatine peak with increase in choline to creatine ratio, decreased N-acetyl aspartate and mildly increased lactate, suggesting high-grade tumor.

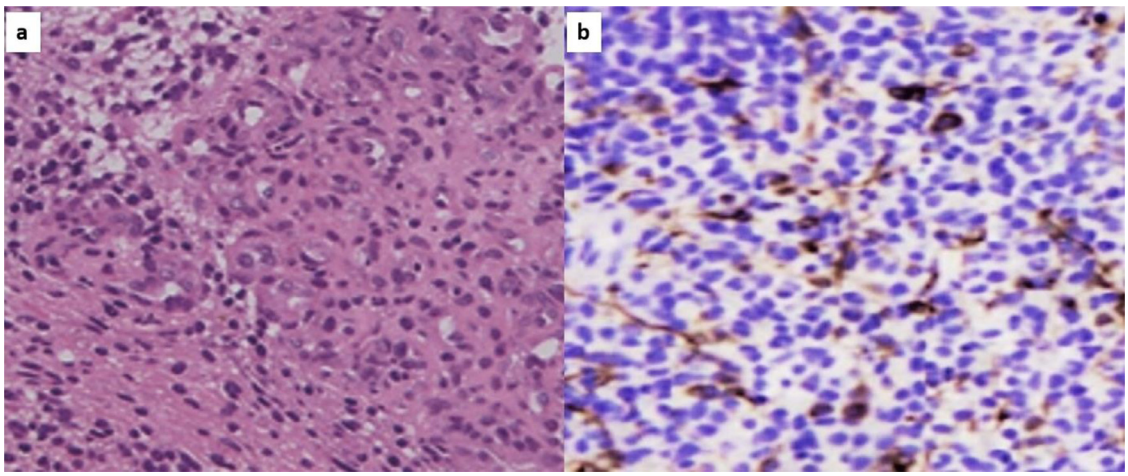

Fig. 5 – Cerebellopontine angle WHO-Grade 4 glioma (glioblastoma): (A) Hematoxylin and eosin (40×) staining shows highly anaplastic glial cells with nuclear atypia, (B) immunohistochemical analysis (40×) reveals positive staining for glial fibrillary acid protein (GFAP) in the tumor cells.

choline to creatine ratio, decrease N-acetyl aspartate and increase choline and creatine peak with mild increase in lactate peak (Fig. 4). The patient underwent near total resection of the tumor and the specimen sent for histopathologic examination.

Histology specimens showed hyperchromatic nuclei and necrosis, which showed positive staining for glial fibrillary acid protein (GFAP) in the tumor cells on immunohistochemical analysis (Fig. 5).

Patient was treated with adjuvant radiotherapy with 60 Gray in 30 fraction at 2Gy/fraction over 6 weeks with concurrent temozolamide (75 mg/m^2).

Discussion

The most common tumor in the CPA is vestibular schwannoma, followed by meningioma, epidermoid cyst, remaining percentage includes different tumors [1]. High-grade glioma (WHO grade4) is common representing 14.6% of central nervous system (CNS) and 48% of malignant brain tumors. Location of glioblastoma in infratentorial location is rare with a frequency of 1.2%-4% cases [2–4]. Incidence of glioblastoma is 10 per 10,000 people globally and is mostly found in the supratentorial region [5]. These tumors are infiltrating, located in the deep white matter or in deep gray matter, mainly in cerebral hemispheres. Such lesion in CPA is even rarer [6,7]. These tumors can develop secondary to diffuse astrocytoma or anaplastic astrocytoma. However, primary glioblastoma can

also be seen [8]. Primary CPA glioblastomas can be classified into 2 variants depending on the site of their origin. The first type of primary glioblastoma is an intra-axial tumor, arising from cerebellum or brain stem with exophytic component into the CPA. The second type is an extra-axial CPA glioblastoma. There have been few cases reported which were arising from the proximal part of cranial nerve VIII [9]. Primary glioma in CPA arises from the root entry zone (either the glial segment or the transition zone) of the cranial nerves in the CPA. There are 3 distinct histological segments of cranial nerves: the glial segment, transition zone, and a peripheral segment. The glial segment is histologically similar to the central nervous system in structure, while peripheral segment structure to a peripheral nerves. There is a transitional or intermediate zone in between glial and peripheral segments [10,11]. Ten cases of primary glioblastoma in CPA have been reported, out of which 6 cases were intra-axial tumors arising from cerebellum or brainstem with exophytic growth into the CPA and 3 were primary extra-axial CPA glioblastoma [7,9,12–16]. In our case, the tumor was separated from the brainstem and was infiltrating the cerebellum with ill-defined margins. There is difficulty in preoperative diagnosis of these CPA glioblastomas including our case. These cases can be misdiagnosed as extra-axial tumors which are more common. The imaging findings in our case suggested meningioma; however, in addition, MRS differentiated malignant tumor from benign. Breshears et al. [12] described glioblastoma as peripherally enhancing cystic mass which was arising from trigeminal nerve root entry zone. On MRI, the lesion was hyperintense on T2-weighted image and hypointense on T1-weighted image with minimal surrounding edema. MRS revealed increased choline with increased N-acetylaspartate [12]. Kasliwal et al. [13] reported multicentric glioblastoma in an 11-year-old child presenting as cerebellopontine angle mass lesion. Panigrahi et al. [14] described CPA glioblastoma in an adult as heterogeneously enhancing lesion in left CPA with no restriction on DWI. Decrease in NAA peak with raised in creatine and choline peak in MRS has been reported, which was also evident in our case. Stark et al. [17] in his study between January 1991 and March 2008 reported 7 infratentorial glioblastoma out of 577 patients with incidence of 1.2%. Hence concluded its rarity in adults with incidence of 1.2% of all patients with glioblastoma [17]. Picart et al. [18] reported case series of 17 adult cerebellar glioblastoma which was compared with supratentorial glioblastoma, suggesting adult cerebellar glioblastoma differs from their supratentorial counterpart. Yang et al. [19] reported a primary glioblastoma of cerebellopontine angle in an adult which was mimicking with preoperative imaging findings of acoustic neuroma. CPA glioblastoma was arising from the proximal portion of VIII cranial nerves. Gopalkrishnan et al. [20] in 2012 did retrospectively analyze the patients with cerebellar glioblastoma over a period of 10 years and reported 5 adults with cerebellar glioblastoma accounting less than 1%. Kuroiwa et al. [21] described posterior fossa glioblastoma as heterogeneous (presence of intratumoral bleed), prominent heterogeneous and ring-like enhancement (necrosis) with poorly defined margins. Zito et al. [22] reported the radiologic findings of posterior fossa glioblastoma, such as lack of peritumoral edema and little mass effect. However, no definite MR findings available to differentiate glioblastoma in CPA from the other benign extra-axial tumors. Doddamani et al. [23] illustrated the characteristics of dural-based glioblastoma in 11 patients describing unique features of this rare presentation, since these tumors can mimic meningioma. In rare cases involvement of dura mater can be seen, which could mimic a meningioma. Transtentorial spread of glioblastoma to cerebellopontine angle has also been reported in the literature by Caramanti et al. [24], which described the metastatic lesion from the primary in left parietal and occipital region. The lesion in CPA was T1-weighted gadolinium enhanced showing cystic areas and central necrosis in right CPA.

Conclusion

Glioblastoma (WHO grade 4) can mimic the imaging features of other benign tumors of CPA. Glioblastoma should be considered in the differential diagnosis of any atypical tumors of CPA. Since clinical manifestations are similar in most of these tumors, hence making an appropriate diagnosis is important for the management of these tumors.

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

REFERENCES

[1] Lalwani AK. Meningiomas, epidermoids, and other nonacoustic tumors of the cerebellopontine angle. Otolaryngol Clin North Am 1992;25:707–28.
[2] Ostrom QT, Cioffi G, Gittleman H, Patil N, Waite K, Kruchko C, et al. CBTRUS statistical report: primary brain and central nervous system tumors diagnosed in the United States in 2012-2016. Neuro Oncol 2019;21:v1–100.
[3] Wu B, Liu W, Zhu H, Feng H, Liu J. Primary glioblastoma of the cerebellopontine angle in adults. J Neurosurg 2011;114:1288–93.
[4] Walter J, Koch A, Herbold C, Schiffler S, Reichart R, Waschke A, et al. Multifocal glioblastoma multiforme in the posterior fossa mimicking cerebral metastases: case presentation and review of the current literature. J Neurol Surg A Cent Eur Neurosurg 2013;74:e30–5.
[5] Hanif F, Muzaffar K, Perveen K, Malhi SM, Simjee SU. Glioblastoma multiforme: a review of its epidemiology and pathogenesis through clinical presentation and treatment. Asian Pac J Cancer Prev 2017;18:3.
[6] Jindal A, Jaiswal AK, Jaiswal S, Pandey P, Mahapatra AK, Sharma MC. Cerebellar glioblastoma multiforme presenting as a cerebellopontine angle mass. J Pediatr Neurosci 2006;1:21–3.
[7] Lee JH, Kim JH, Kwon TH. Primary glioblastoma of the cerebellopontine angle: case report and review of the literature. J Korean Neurosurg Soc 2017;60:380.
[8] Vanderberg SR. Current diagnostic concepts of astrocytic tumors. J Neuropathol Exp Neurol 1992;51:644–57.

[9] Mariniello G, Peca C, de Caro M, Corvino S, Orlando V. Cerebello-pontine angle glioblastoma with cervical spine metastasis: a case report. Neurosurg Cases Rev 2019;2:12.
[10] Arnautovic KI, Husain MM, Linskey ME. Cranial nerve root entry zone primary cerebellopontine angle gliomas: a rare and poorly recognized subset of extraparenchymal tumors. J Neuro-Oncol 2000;49:205–12. doi:10.1023/a:1006488905526.
[11] Francesco F, Maurizio I, Stefano C, Marina S, Ugo S, Massimo S. Trigeminal nerve root entry zone pilocytic astrocytoma in an adult: a rare case of an extraparenchymal tumor. J Neuro-Oncol 2010;97:285–90. doi:10.1007/s11060-009-0020-4.
[12] Breshears JD, Ivan ME, Cotter JA, Bollen AW, Theodosopoulos PV, Berger MS. Primary glioblastoma of the trigeminal nerve root entry zone: case report. J Neurosurg 2015;122:78–81. doi:10.3171/2014.10.JNS14449.
[13] Kasliwal MK, Gupta DK, Mahapatra AK, Sharma MC. Multicentric cerebellopontine angle glioblastoma multiforme. Pediatr Neurosurg 2008;44:224–8. doi:10.1159/000121380.
[14] Panigrahi S, Mishra SS, Das S. Primary cerebellopontine angle glioblastoma in an adult. Asian J Neurosurg 2017;12(1):62–4. doi:10.4103/1793-5482.145323.
[15] Matsuda M, Onuma K, Satomi K, Nakai K, Yamamoto T, Matsumura A. Exophytic cerebellar glioblastoma in the cerebellopontine angle: case report and review of the literature. J Neurol Surg Rep 2014;75:e67–72.
[16] Salunke P, Sura S, Tewari MK, Gupta K, Khandelwal NK. An exophytic brain stem glioblastoma in an elderly presenting as a cerebellopontine angle syndrome. Br J Neurosurg 2012;26:96–8.
[17] Stark AM, Maslehaty H, Hugo HH, Mahvash M, Mehdorn HM. Glioblastoma of the cerebellum and brainstem. J Clin Neurosci 2010;17(10):1248–51. doi:10.1016/j.jocn.2010.02.015.
[18] Picart T, Barritault M, Berthillier J, Meyronet D, Vasiljevic A, Frappaz D, et al. Characteristics of cerebellar glioblastomas in adults. J Neurooncol 2018;136(3):555–63. doi:10.1007/s11060-017-2682-7.
[19] Yang DX, Jing Y, Xu ZM, Yuan F, Liu YL, Wang GS, et al. Primary glioblastoma of cerebellopontine angle in adult mimicking acoustic neuroma. World Neurosurg 2019;122:48–52. doi:10.1016/j.wneu.2018.10.073.
[20] Gopalakrishnan CV, Dhakoji A, Nair S, Menon G, Neelima R. A retrospective study of primary cerebellar glioblastoma multiforme in adults. J Clin Neurosci 2012;19(12):1684–8. doi:10.1016/j.jocn.2011.12.035.
[21] Kuroiwa T, Numaguchi Y, Rothman MI, et al. Posterior fossa glioblastoma multiforme: MR findings. AJNR Am J Neuroradiol 1995;16(3):583–9.
[22] Zito JL, Siva A, Smith TW, Leeds M, Davidson R. Glioblastoma of the cerebellum. Computed tomographic and pathologic considerations. Surg Neurol 1983;19(4):373–8.
[23] Doddamani RS, Meena RK, Sawarkar D. Ambiguity in the dural tail sign on MRI. Surg Neurol Int 2018;9:62.
[24] Caramanti RL, Aprígio RM, Tognola WA, Laurenti MR, Rocha CE, Góes MJ. Transtentorial spread of glioblastoma multiforme to cerebellopontine angle—a rare case report. Surg Neurol Int 2022;13:5. doi:10.25259/SNI_948_2021.