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Case Report

Extra axial medulloblastoma of the cerebellopontine angle: A rare case report☆,☆☆

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ABSTRACT

Medulloblastoma is the most frequent malignant brain tumor in children. Originating in the cerebellum, they are typically intra-axial tumors. In adults, they represent less than 1% of brain tumors. However, the occurrence of extra-axial medulloblastoma is possible but extremely rare, and slightly more frequent in the adult population. We present a rare case of extra-axial medulloblastoma, diagnosed in a 22-year-old male, through advanced imaging techniques, followed by confirmation through anatomopathological examination.
This case calls attention to the necessity of knowledge of the various diagnostic possibilities when interpreting radiological images, leading to enhanced patient care and furthering our understanding of these exceptional entities.

Introduction

Medulloblastoma is a high-grade malignant tumor classified as an embryonal neuroepithelial tumor [1]. It represents 15%-20% of central nervous system tumors in individuals under the age of 15 and accounts for one-third of posterior fossa tumors [2]. Typically, medulloblastomas originate from the cerebellar vermis in childhood, while in adulthood, they arise from the paramedian or lateral regions within the cerebellar hemisphere, where they represent less than 1% of primary brain tumors [3]. However, a few cases of the posterior fossa extra-axial variant of medulloblastomas have been reported in the literature, most of which are located in the cerebello-pontine angle (CPA) [4]. This raises the issue of differential diagnosis with common tumors of the CPA, such as vestibular

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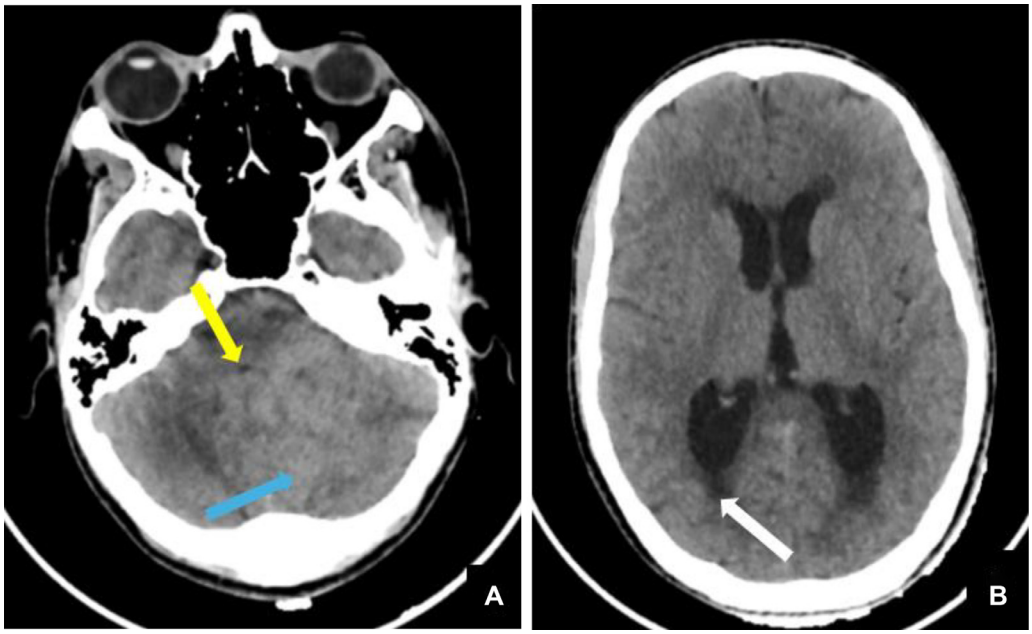

Fig. 1 – Precontrast CT scan reveals the presence of an expansive solid mass of the posterior cerebral fossa (blue arrow) with mass effect on the fourth ventricle (yellow arrow) (A). Trans ependymal resorption (white arrow) (B).

schwannomas, meningiomas, and epidermoid inclusion cysts [5].

We report a case of CPA medulloblastoma in a 22-year-old male who was surgically treated, highlighting that an extra-axial mass in the CPA can indeed be a medulloblastoma.

Case presentation

A 22-year-old male patient with no significant medical history was admitted to the emergency department after experiencing several episodes of vomiting and headaches over the past 2 weeks. During the subsequent neurological assessment, gait disturbances and ataxia were noted over the last 3 weeks. No other neurological disorders were present.

A cranial computed tomography (CT) scan was ordered. The CT scan, performed using a 64-slice GE scanner without contrast administration, revealed a solid, spontaneously hyperdense mass in the left posterior fossa. This mass caused a significant effect on the fourth ventricle, resulting in triventricular obstructive hydrocephalus with periventricular hypodensity consistent with trans-ependymal oedema (Fig. 1).

For further characterization of the lesion, a brain MRI was performed, revealing a well-defined voluminous extra-axial mass in the left cerebellopontine angle (CPA), measuring 37 × 48 × 30 mm. The mass exhibited a hypointense signal on T1-weighted images, isointense signal on FLAIR imaging, and slightly hyperintense signal on T2-weighted images. It was surrounded by a thin layer of cerebrospinal fluid (CSF), confirming its extra-axial origin (Fig. 2).

Diffusion-weighted imaging showed hyperintensity with clear restriction of diffusion (low ADC values). Following the administration of a gadolinium-based contrast agent, the mass exhibited homogeneous enhancement (Fig. 3). Notably, there was pronounced vasogenic edema adjacent to the tumor, evident as a high-intensity signal on FLAIR imaging, and also in the periventricular region (Fig. 4).

The tumor exerted a mass effect on the right cerebellar hemisphere, causing inferior herniation of the right cerebellar tonsil, as well as effacement of the fourth ventricle, resulting in obstructive hydrocephalus (Fig. 5). An MRI of the spinal cord was performed, revealing multiple extramedullary nodular lesions at various cervical, thoracic, and lumbar levels that showed significant enhancement following gadolinium injection (Fig. 6).

The patient underwent surgical resection of the tumor due to the mass effect and hydrocephalus, and histopathological analysis revealed extra-axial medulloblastoma.

Discussion

Medulloblastoma represents the most common malignant tumor of the posterior cerebral fossa in childhood, accounting for about 40% of cases [6]. The majority of medulloblastomas arise in children, with a median age of diagnosis at 9 years; a second peak is observed in adults, accounting for around 25% of cases [7]. Typically, these tumors are intra-axial, with 80% arising in the cerebellar midline at the inferior vermis, often projecting into and filling the fourth ventricle [8]. A smaller proportion of medulloblastomas are located in the cerebellar hemispheres, usually manifesting in adolescents or young adults.

The origin of medulloblastoma is believed to be linked to germinal cells or their remnants located at the inferior medullary velum or to remnants of the external granular layer. However, the precise origin of this tumor remains uncertain [9].

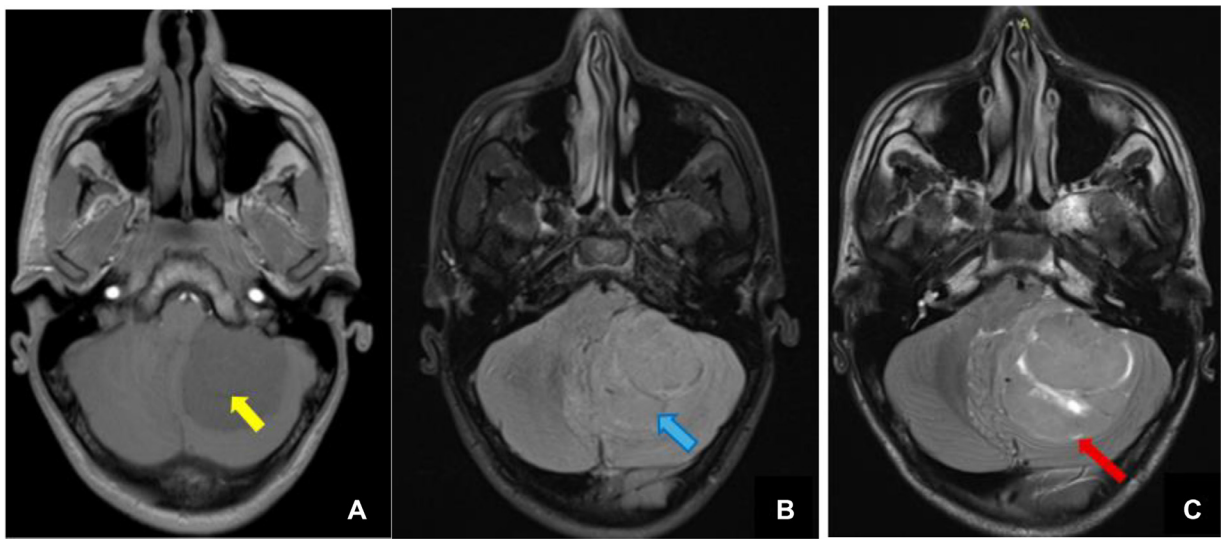

Fig. 2 – Hypo intense signal on T1 (A) (yellow arrow), iso signal on FLAIR (B) (blue arrow) and a thin layer of CSF surrounding tumor on T2 (red arrow).

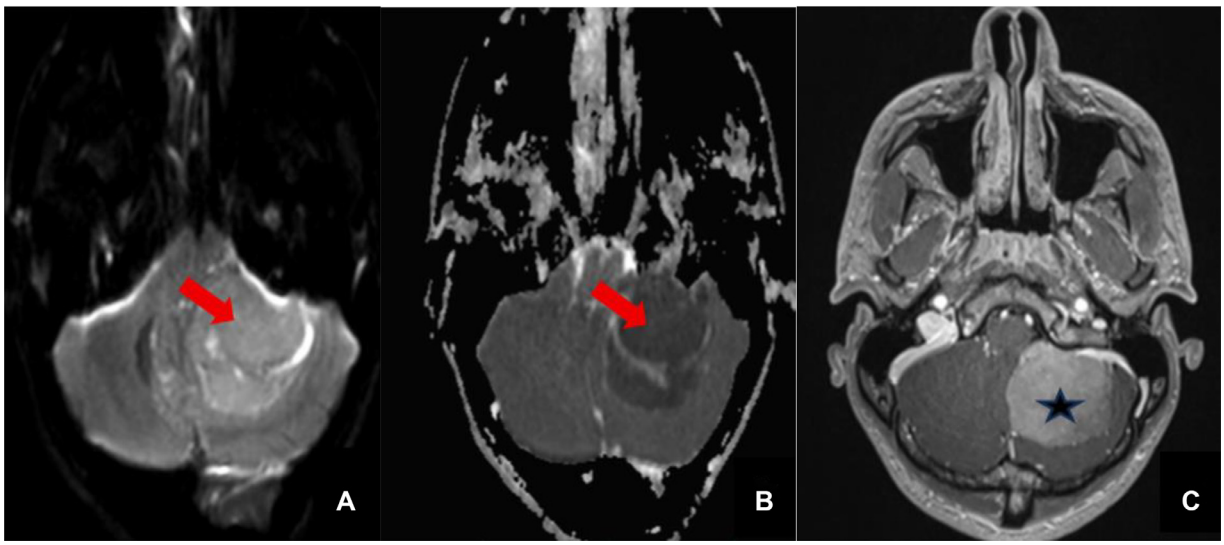

Fig. 3 – Hypersignal on diffusion-weighted imaging (A), low ADC value (B) (red arrows), homogeneous enhancement (C) (stars).

The classification of medulloblastoma includes 4 molecular subgroups: WNT, SHH, Group 3, and Group 4. The WNT subgroup, characterized by classic features, has an excellent prognosis. The SHH subgroup can present as desmoplastic or classic and shows variable prognoses, with the desmoplastic variant having more favorable outcomes. Group 3 is associated with a poor prognosis, while Group 4 presents an intermediate prognosis. Notably, except for the SHH subgroup, which primarily manifests in infancy or adulthood, the other subgroups predominantly occur in childhood.

Extra-axial medulloblastoma remains an extremely rare location for this tumor; it predominantly occurs in the cerebellopontine angle (CPA), with only 42 cases reported in the literature [10].

In our case, the initial diagnosis involved confirming the extra-axial nature of the tumor, supported by several discernible signs. The most apparent indicator was the presence of the cerebrospinal fluid (CSF) cleft sign, clearly visualized on FLAIR imaging, indicating that CSF serves as a barrier between the lesion and brain parenchyma. The absence of significant edema further supports its extra-axial localization; intra-axial lesions typically exhibit significant edema.

In the context of CPA tumors, there are several potential differential diagnoses to consider. Vestibular schwannoma is the most common nonmalignant nerve sheath tumor, accounting for 90% of cerebellopontine angle tumors [11]. When associated with medullary schwannoma, it is highly suggestive of neurofibromatosis type 2 (NF2), which was our first radi-

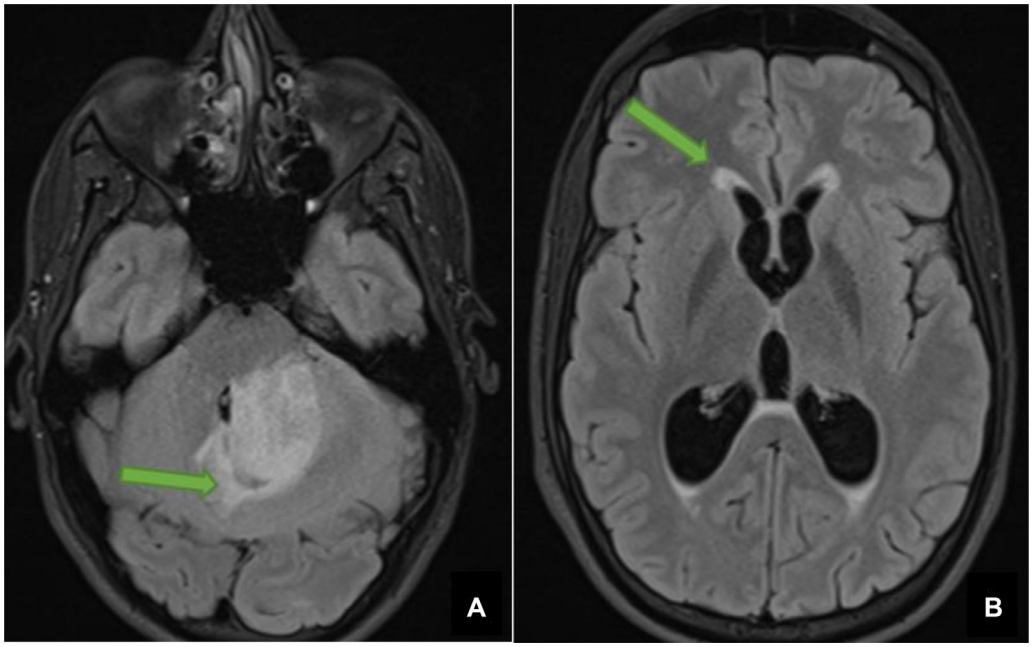

Fig. 4 – vasogenic oedema adjacent to the tumor (A), and on periventricular (B) (green arrows).

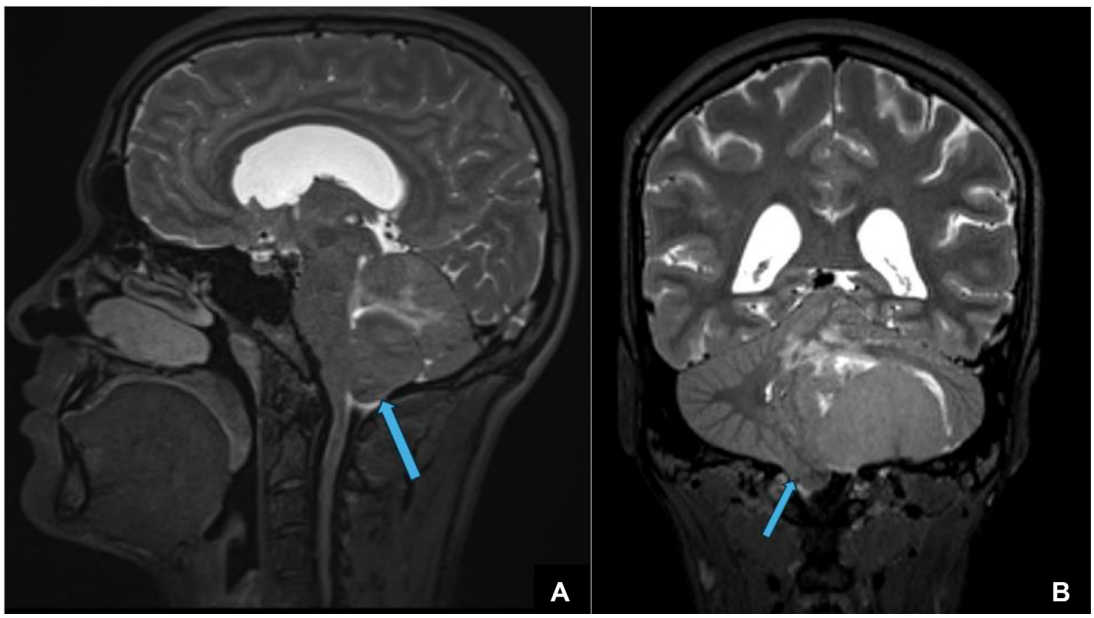

Fig. 5 – (A) Sagittal, (B) Coronal: herniation of the right cerebellar tonsil (blue arrows).

ological diagnosis. Vestibular schwannomas typically present as slightly hypointense on T1-weighted images and heterogeneously hyperintense on T2-weighted images, with homogeneous contrast enhancement after gadolinium injection. Some tumors may show heterogeneous contrast enhancement due to cystic changes [12]. Additional differential diagnoses include meningioma, primary cholesteatomas, and epidermoid tumors.

Differentiating extra-axial medulloblastoma from other tumors in the CPA can be challenging. Clinical signs of intra-axial lesions may include brainstem and cerebellar dysfunction that could suggest medulloblastomas [13]. They typically exhibit rapidly progressive evolution and are less common to present with cranial nerve involvement [14].

Radiologically, extra-axial medulloblastoma is characterized by a hypointense signal on T1-weighted images with homogeneous or heterogeneous contrast enhancement and hyperintensity on T2-weighted images. Restricted diffusion on DWI and low ADC values are also noted.

Spinal metastasis from CPA medulloblastoma has been reported in only one patient previously [15], our case represents the second instance.

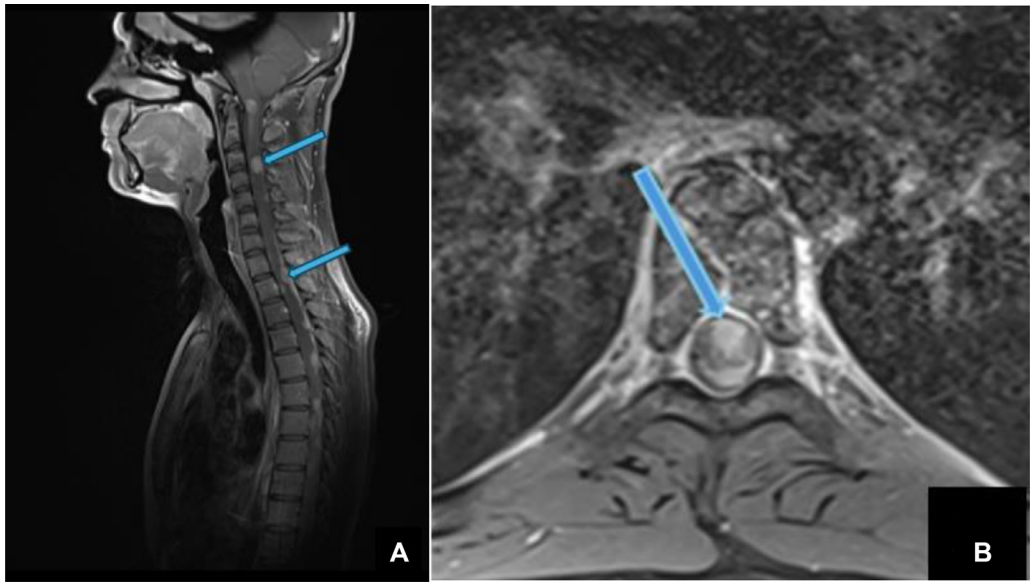

Fig. 6 – (A) Sagittal, (B) Axial: multiple extramedullary nodular lesions (blue arrows).

The treatment of extra-axial medulloblastoma remains controversial due to limited reported cases. Surgery with adjuvant radiotherapy is commonly performed as the main treatment modality [16]. Chemotherapy has also been combined in some cases.

A 5-year survival rate of approximately 30% has been reported for CPA medulloblastoma, which is comparable to that of medulloblastoma located in other areas [13].

Conclusion

In summary, while medulloblastoma is typically an intra-axial pediatric tumor, extra-axial presentations, particularly in the cerebellopontine angle (CPA) of adults, are exceedingly rare. Nevertheless, when evaluating extra-axial CPA lesions especially those with atypical radiological features or rapid progression, medulloblastoma should be included in the differential diagnosis to ensure appropriate management and improve patient outcomes. Current literature indicates that only a limited number of cases have been reported, highlighting the necessity for heightened awareness among clinicians regarding this unusual manifestation of medulloblastoma in adults.

Patient consent

Complete written informed consent was obtained from the patient for the publication of this study and accompanying images.

REFERENCES

[1] Mahapatra S, Amsbaugh MJ. Medulloblastoma. StatPearls. Treasure IslandFL: StatPearls Publishing; 2023.

[2] Tumeurs cérébrales. du diagnostic au traitement. Masson, Paris: J. Philippon; 2004. ISBN 2-294-01288-7 p. 285.

[3] Menon G, Krishnakumar K, Nair S. Adult medulloblastoma: clinical profile and treatment results of 18 patients. J Clin Neurosci 2008;15:122–6.

[4] Al-Sharydah AM, Al-Abdulwahhab AH, Al-Suhibani SS, et al. Posterior fossa extra-axial variations of medulloblastoma: a pictorial review as a primer for radiologists. Insights Imaging 2021;12:43. doi:10.1186/s13244-021-00981-z.

[5] Akay KM, Erdogan E, Izci Y, Kaya A, Timurkaynak E. Medulloblastoma of the cerebellopontine angle. Neurol Med Chir 2003;43:555–8.

[6] Thanh Dung L, Minh Duc N. Medulloblastoma in the cerebellopontine angle mimicking a schwannoma. Clin Case Rep 2021;9(4):1948–53. doi:10.1002/ccr3.3912.

[7] Orr BA. Pathology, diagnostics, and classification of medulloblastoma. Brain Pathol (Zurich, Switzerland) 2020;30(3):664–78. doi:10.1111/bpa.12837.

[8] Pant I, Chaturvedi S, Gautam VK, Pandey P, Kumari R. Extra-axial medulloblastoma in the cerebellopontine angle: report of a rare entity with review of literature. J Pediatr Neurosci 2016;11(4):331–4. doi:10.4103/1817-1745.199477.

[9] Bahrami E, Bakhti S, Fereshtehnejad SM, Parvaresh M, Khani MR. Extra-axial medulloblastoma in cerebello-pontine angle: a report of a rare case with literature review. Med J Islam Repub Iran 2014;28:57.

[10] Balasubramanian K, Kharbat AF, Call-Orellana F, Tavakol SA, Fassina GR, Janssen C, et al. Adult Cerebellopontine angle Medulloblastoma: a systematic review of clinical features, management approaches, and patient outcomes. Cancers 2024;16(24):4242. doi:10.3390/cancers16244242.

[11] Cioffi G, Yeboa DN, Kelly M, Patil N, Manzoor N, Greppin K, et al. Epidemiology of vestibular schwannoma in the United States, 2004–2016. Neuro-Oncol Adv 2020;2(1):vdaa135. doi:10.1093/noajnl/vdaa135.

[12] Nguyen D, de Kanztow L. Vestibular schwannomas: a review. Appl Rad Oncol 2019;48(3):22–7.

[13] Noiphithak R, Yindeedej V, Thamwongskul C. Cerebellopontine angle medulloblastoma with extensive nodularity in a child: case report and review of the literature. Child's Nerv Syst 2017;33(5):839–42. doi:10.1007/s00381-016-3325-6.

[14] Bonneville F, Sarrazin J-L, Marsot-Dupuch K, et al. Unusual lesions of the cerebellopontine angle: a segmental approach. Radiographics 2001;21:419–38. doi:10.1148/radiographics.21.2.g01mr13419.
[15] Ebrahimzadeh K, Mirahmadi Eraghi M, Tavassol H H, Hallajnejad M, Rezaei Mirghaed O, Naghibi Irvani S, et al. The extra-axial cerebellopontine angle medulloblastoma in an adult patient: a case report and review of literature. Iran J Neurosurg 2022;8(1):32. URL: http://irjns.org/article-1-303-en.html.
[16] Kumar R, Bhowmick U, Kalra S, Mahapatra A. Pediatric cerebellopontine angle medulloblastomas. J Pediatr Neurosci 2008;3:127. doi:10.4103/1817-1745.43638.