

Bifocal medulloblastoma in an adult: A case report and review of the literature

Ghizlane Lembarki *, Otmane Soussi, Mouna Sabiri, Chorouk Mountassir, Mohamed Labied and Samira Lezar

Central Unit of Radiology, Ibn Rochd University Hospital, Casablanca, Morocco.

World Journal of Biology Pharmacy and Health Sciences, 2024, 19(02), 107–111

Publication history: Received on 23 June 2024; revised on 02 August 2024; accepted on 04 August 2024

Article DOI: <https://doi.org/10.30574/wjbphs.2024.19.2.0487>

Abstract

Medulloblastoma is a malignant embryonal neuroepithelial tumour, rarely occurring in adults. The 2021 WHO CNS classification subdivided medulloblastoma into 4 groups based on molecular data, which may correspond to morphological features on imaging which are useful to recognise. Differential diagnoses in adults include ependymoma, hemangioblastoma, metastases, lymphoma, and high-grade glioma.

We report the case of a double cerebellar medulloblastoma in a 34-year-old adult. We also highlight the differential diagnosis between medulloblastoma and other posterior cerebellar fossa tumours in adults providing practical elements to help suggest the most accurate diagnosis.

Keywords: Adults medulloblastoma; CNS WHO 2021 classification; CT; MRI; Differential diagnosis.

1. Introduction

Medulloblastoma occurs predominantly in children, with a peak age of 5 years and a slight male predominance. Its occurrence in adults remains rare, but should always be suspected on imaging.

We report a case of a double cerebellar medulloblastoma occurring in a 34-year-old adult.

2. Case report

A 34-year-old male, without a noticeable medical history, presented with ataxia and intracranial hypertension syndrome that developed over a month, accompanied by a decline in overall health.

A brain CT scan revealed two intra-axial, bilateral rounded cerebellar lesions, quite well limited, isodense, with discreet enhancement after contrast media agent administration. These lesions compressed the fourth ventricle, resulting in an active triventricular hydrocephalus (figure 1). These two lesions appeared on MRI as hypointense T1, discretely hyperintense T2 lesions with some cystic areas, diffusion restriction and discrete enhancement after gadolinium-containing contrast agent administration (figure 2).

* Corresponding author: Ghizlane Lembarki.

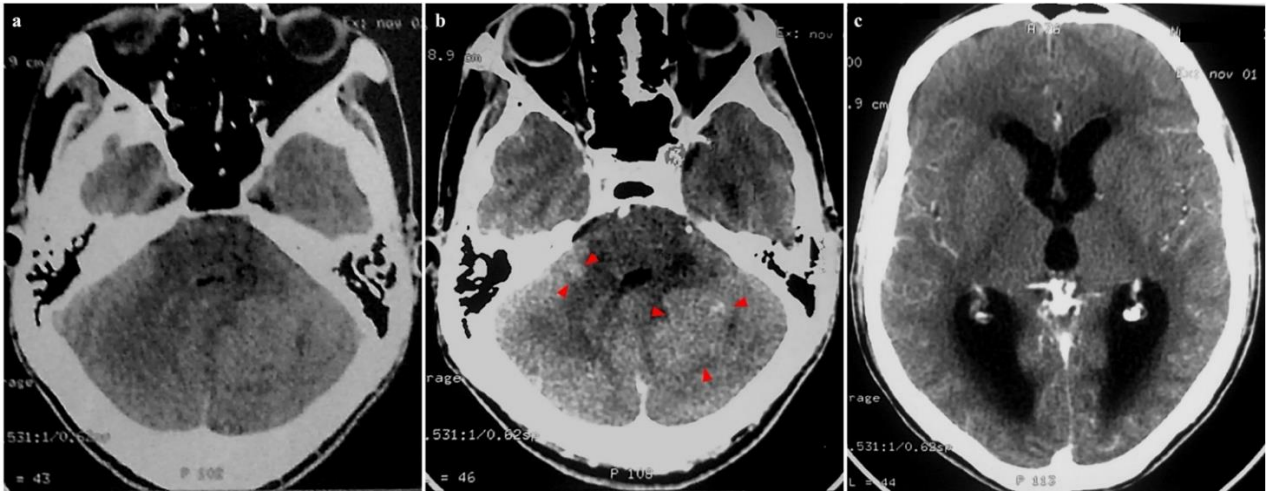


Figure 1 Brain CT scan in axial sections before (a) and after administration of contrast media agent (b, c), showing two isodense hemispheric cerebellar processes, discretely enhanced (red arrowhead), and surrounded by oedema. They have a mass effect on the 4th ventricle, which is responsible for a triventricular hydrocephalus.

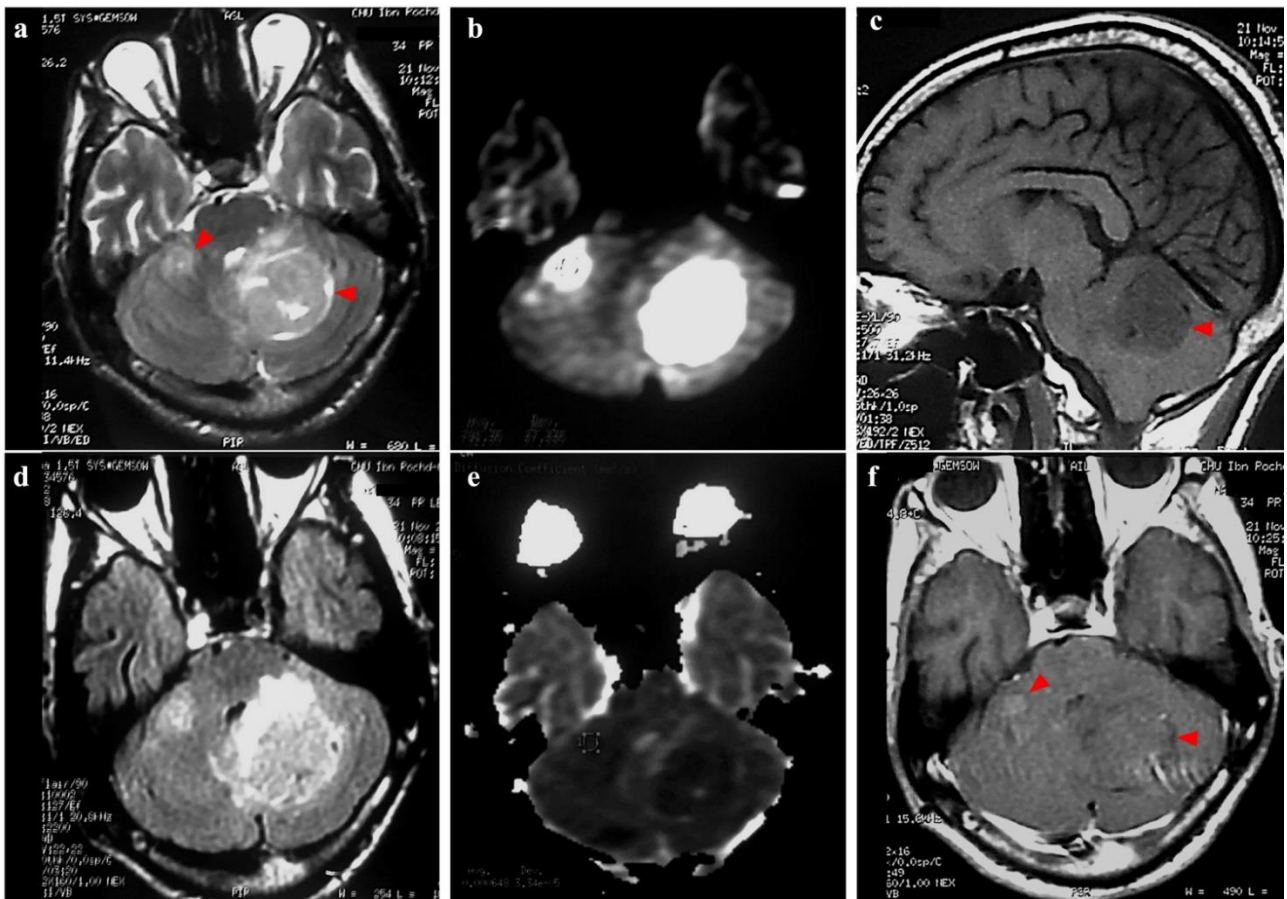


Figure 2 Brain MRI in the same patient, showing the cerebellar processes (red arrowhead) in hypersignal T2 and FLAIR (a, d), hypersignal diffusion with very low ADC (b, e), hyposignal T1 (c), discretely enhanced after gadolinium containing contrast agent administration (f).

After stereotaxic biopsy, pathology revealed a malignant tumour proliferation, formed by small round cells arranged in diffuse sheets with pseudorosettes and nodular foci: these findings were consistent with a classic medulloblastoma.

3. Discussion

Medulloblastoma is an undifferentiated neuroepithelial tumour, characterised by its aggressive nature, and its prevalence frequently in children: it affects children under the age of 15 in 80% of cases, with a peak prevalence at age 5. However, medulloblastoma can occur in adults (around 30 years old), accounting for 1% of brain tumours in this age range. [1]

The new WHO 2021 classification of CNS tumours has classified medulloblastomas into two categories: histologically diagnosed medulloblastomas (classic, desmoplastic/nodular, medulloblastoma with extensive nodularity (MBEN), and large cell/anaplastic) and molecularly diagnosed medulloblastomas (WNT-activated, SHH-activated, non-WNT/non-SHH: group 3 and group 4) that give a better prediction of clinical behaviour and outcome [2, 5]. However, there is a correlation between these two items: desmoplastic/nodular medulloblastomas and MBEN are in the SHH molecular group, 94% of WNT tumours have a classic morphology, and most large cell/anaplastic tumours belong either to the SHH-3 subgroup or to non-WNT/non-SHH group. [2, 3]

Various studies report that the lateralized location of medulloblastoma is more frequent in adults than in children. In clinical terms, this cerebellar hemisphere localization would result in a kinetic cerebellar syndrome and a vestibular syndrome, rather than a static cerebellar syndrome. In adults, reports of intracranial hypertension syndrome are less common and are revealed later than in children [1, 4]

Medulloblastoma is a tumour that is very prone to dissemination through the CSF, which could explain secondary cerebral, medullary, or leptomeningeal locations; in this case, they manifest in the form of nodular lesions, preferably peripheral in location [4], and should be systematically investigated with gadolinium-enhanced T1 MRI of the brain and the entire spinal canal. [5]

In adults, it appears as a hyperdense mass on a CT scan that is lateralized and has a well-defined margin. It may also include cystic or necrotic regions and calcifications. In MRI, it appears as hypointense T1, iso or hyperintense T2/FLAIR, diffusion hyperintense with low ADC values. Whether in CT or MRI, this tumour shows discrete homogeneous or heterogeneous enhancement after contrast media administration. [4]

Some studies have shown that morphological features in imaging strongly orient towards specific groups: medulloblastomas centred on the cerebellar peduncles often belong to the WNT-activated group, which has a good prognosis. Hemispheric location is often associated with the SHH-activated group, which has a poor prognosis. A medial location associated with minimal enhancement suggests a group 3 or 4 medulloblastoma. [6]

The differential diagnosis of medulloblastoma in adults includes ependymoma, hemangioblastoma, metastases, lymphoma, and high-grade glioma. Ependymoma is the primary differential diagnosis, which is frequently confused with medulloblastoma. However, many studies have shown distinguishing features: medulloblastoma in adults arises around the age of 30 (earlier than ependymoma), is frequently lateralized to the cerebellar hemispheres, has a low ADC, and has little or no enhancement after contrast agent administration [6]. Table 1 shows the main clinical and radiological characteristics of medulloblastoma and its differential diagnoses. [5, 7]

Table 1 Main clinical and radiological features of adult medulloblastoma and its differential diagnoses.

	Medulloblastoma	Ependymoma	Hemangioblastoma	Metastases	Lymphoma	High-grade glioma	
Average age	Adult ≈ 30 years	Adult > 50 years	Adult ≈ 30 years	Without peak	Adult ≈ 50 years	Adult ≈ 50 years	
Location	cerebellar hemisphere	In the 4th ventricle	Hemisphere: 2/3 of case Vermis: 1/3 of case	Variable, multiples++	Periventricular	Brain stem Cerebellum	
CT scan	hyperdense	Heterogeneous Iso or hyperdense	Large cystic component with a mural nodule	Iso or hypodense	Iso or hyperdense	Heterogeneous Hypodense (hemorrhage and necrosis)	
MRI	T1	hypointense	Isosignal	Isosignal	Iso, hypo or hypersignal (depends on the primary lesion)	Iso or hyposignal	Heterogeneous hyposignal
	T2	iso or hyperintense	Hypersignal	Hypersignal	Discrete hypersignal	Iso or hypersignal	Hypersignal
	Diffusion	hyperintense with low ADC	Hyposignal with high ADC	Hyposignal with high ADC	Hypersignal with low ADC	Hypersignal with low ADC	Hypersignal with low ADC
	Contrast enhancement	Discrete or no enhancement	Intense enhancement of mural nodule	Intense enhancement of mural nodule	Variable	Intense and homogeneous (heterogeneous in immunocompromised)	Heterogeneous (nodular or annular)
Oedema	Present	discrete	Often absent	Marked	Marked	Marked	
Cyst	Small areas	Small areas	Predominantly	Present: necrosis	Unusual	Variable	
Calcification	Rare	Nodular (>50%)	Absent	Rare	unusual	curvilinear clods	

4. Conclusion

Adult medulloblastoma has a different appearance than childhood medulloblastoma, with a better response to treatment and prognosis. This diagnosis should be considered in adults without forgetting other differential diagnoses.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest is to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

References

- [1] Menon G, Krishnakumar K, Nair S. Adult medulloblastoma: clinical profile and treatment results of 18 patients. *J Clin Neurosci*. 2008 Feb;15(2):122-6. doi: 10.1016/j.jocn.2007.06.007. Epub 2007 Dec 19. PMID: 18078755.
- [2] Louis DN, Perry A, Wesseling P, Brat DJ, Cree IA, Figarella-Branger D, Hawkins C, Ng HK, Pfister SM, Reifenberger G, Soffietti R, von Deimling A, Ellison DW. The 2021 WHO Classification of Tumors of the Central Nervous System: A Summary. *Neuro Oncol*. 2021 Aug 2;23(8):1231-1251. doi: 10.1093/neuonc/noab106. PMID: 34185076; PMCID: PMC8328013.
- [3] Ellison DW, Dalton J, Kocak M, Nicholson SL, Fraga C, Neale G, Kenney AM, Brat DJ, Perry A, Yong WH, Taylor RE, Bailey S, Clifford SC, Gilbertson RJ. Medulloblastoma: clinicopathological correlates of SHH, WNT, and non-SHH/WNT molecular subgroups. *Acta Neuropathol*. 2011 Mar;121(3):381-96. doi: 10.1007/s00401-011-0800-8. Epub 2011 Jan 26. PMID: 21267586; PMCID: PMC3519926.
- [4] Drissi J, Affane M, Elomrani A, Khouchani M. Le médulloblastome chez l'adulte : à propos de 13 cas et revue de la littérature [Medulloblastoma in adults: report of 13 cases and literature review]. *Pan Afr Med J*. 2015 Oct 12;22:126. French. doi: 10.11604/pamj.2015.22.126.7242. PMID: 26889307; PMCID: PMC4742019.
- [5] Dietemann J l., Bogorin A, Eid MA, Koob M. Tumeurs crânio-encéphaliques : tumeurs intra-axiales. Elsevier eBooks [Internet]. 2018. p. 203-296.e15. available on: <https://www.sciencedirect.com/science/article/abs/pii/B9782294753947000072>.
- [6] Perreault S, Ramaswamy V, Achrol AS, Chao K, Liu TT, Shih D, Remke M, Schubert S, Bouffet E, Fisher PG, Partap S, Vogel H, Taylor MD, Cho YJ, Yeom KW. MRI surrogates for molecular subgroups of medulloblastoma. *AJNR Am J Neuroradiol*. 2014 Jul;35(7):1263-9. doi: 10.3174/ajnr.A3990. Epub 2014 May 15. PMID: 24831600; PMCID: PMC4819007.
- [7] Deng J, Xue C, Liu X, Li S, Zhou J. Differentiating between adult intracranial medulloblastoma and ependymoma using MRI. *Clin Radiol*. 2023 Mar;78(3):e288-e293. doi: 10.1016/j.crad.2022.11.016. Epub 2023 Jan 2. PMID: 36646528.