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Case report

Cerbellar medulloblastoma in the elderly: Case report and review of the literature

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ABSTRACT

Introduction: Medulloblastoma is a malignant tumor of the posterior fossa which is mainly found in children aged less than 15 years of age. This tumor is uncommon in adults, especially those above 40 years old. We report the very rare case of a 61 years-old woman who was diagnosed with a cerebellar medulloblastoma.

Case presentation: We present a case of a 61-year-old woman who presented for signs of elevated intracranial and cerebellar syndrome related to an atypical mass located within the left cerebellar hemisphere. The patient underwent an incomplete resection of the mass due to the infiltration of the superior cerebellar peduncle. Histology and immunohistochemistry concluded to a classic medulloblastoma. Thus, she underwent postoperative radiotherapy.

Discussion: Medulloblastoma is very rarely diagnosed in patients above 60 years, as only 14 cases have already been published. Therapeutic approach is based on surgery and radiotherapy in both children and adults, whereas addition of chemotherapy is controversial.

Conclusions: Even in adults over 60 years of age, medulloblastoma should be included in the differential diagnosis of a cerebellar mass. This diagnosis should be kept in mind as clinical and radiological present specificities to the features commonly described in infants' medulloblastoma.

1. Introduction

Medulloblastomas (MB) are known to be the most frequent primitive tumor of the posterior fossa in the childhood. They represent between 20 and 25% of central nervous system (CNS) tumors in the pediatric population [1]. MB are very rarely reported during the adulthood, with approximately 80% of affected adults younger than 35 old [2]. To the best of our knowledge, only 14 cases of late adults diagnosed with a MB above the age of 60 years-old have been reported in the literature [2]. Hereby we report the new case of a 61 years-old patient operated for a cerebellar tumor, whose pathologic and immunochemistry exams revealed a MB.

The work has been reported in line with the SCARE 2020 criteria [3].

2. Case description

We report the case of a 61 years old woman, known to be suffering from arterial hypertension and diabetes, who presented for the progressive onset since 1 month of headaches associated to episodes of vomiting, and dizziness. Physical examination found a fully conscious patient, who had static and left kinetic cerebellar syndromes. The rest of the exam showed neither signs of cranial nerves impairment, nor elements that may orientate towards an extra-cranial tumor. Brain MRI (Figs. 1 and 2) showed two intra-axial lesions within the left cerebellar hemisphere, measuring both 3 cm of diameter, extending towards the left latero-mesencephalic cistern, and probably infiltrating the left superior cerebellar peduncle. These lesions had the same semiologic characters: hypointense on T1-weighted imaging (WI), heterogenous on T2-WI, with a slight enhancement following injection of Gadolinium delimitating cystic intra-tumoral areas. This lesion was hyperintense on diffusion-WI, with a restriction on ADC cartography witnessing their hypercellularity. Perfusion did not show any neoangiogenesis, and spectroscopy objectified a tumoral profile related to a peak of choline and lipids, a NAA drop, and an elevated choline/creat ratio. A slight perilesional edema could be seen on T2 Flair-WI. The tumor was not associated to a mass effect towards ventricular structures, thus no associated hydrocephalus.

These radiologic features were suggestive for left cerebellar

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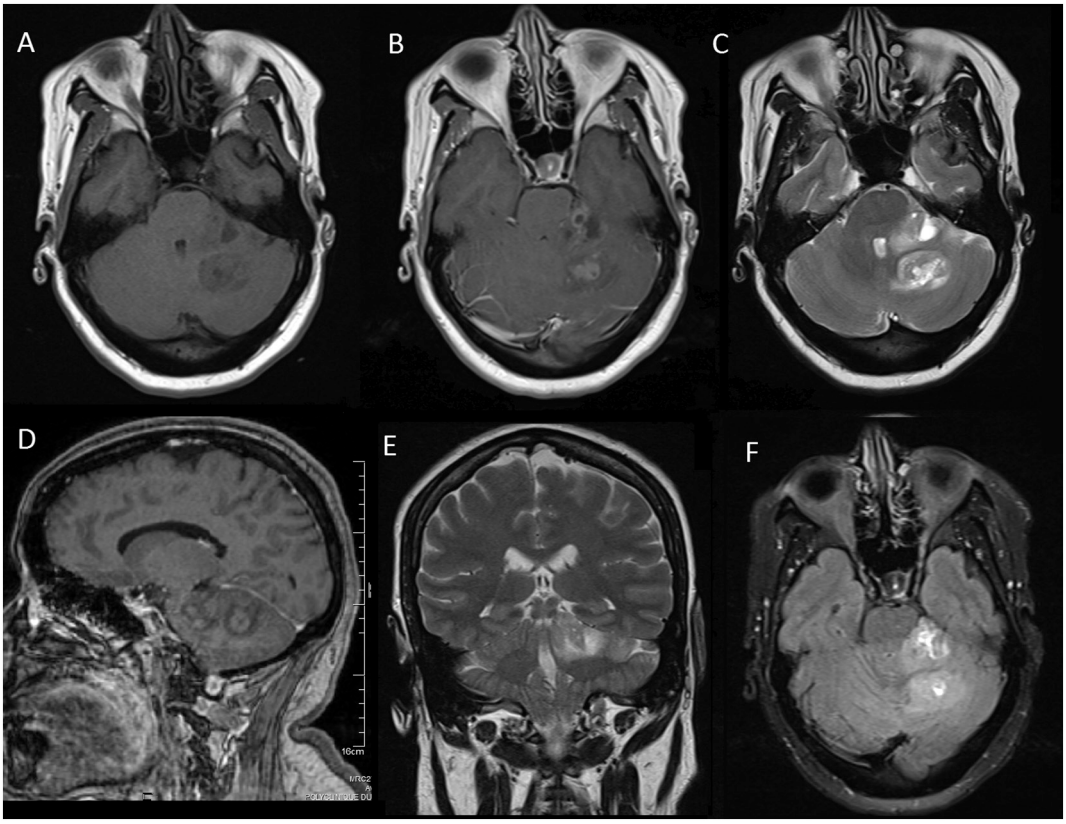

Fig. 1. Axial (A, B, C and F), sagittal (D) and coronal (E) sections of a brain MRI on T1-weighted imaging (WI) without injection of Gadolinium (A), T1-WI with injection of Gadolinium (B and D), T2-WI, (C and E) and T2 flair (E) showing a lesion developing in the left cerebellar hemisphere.

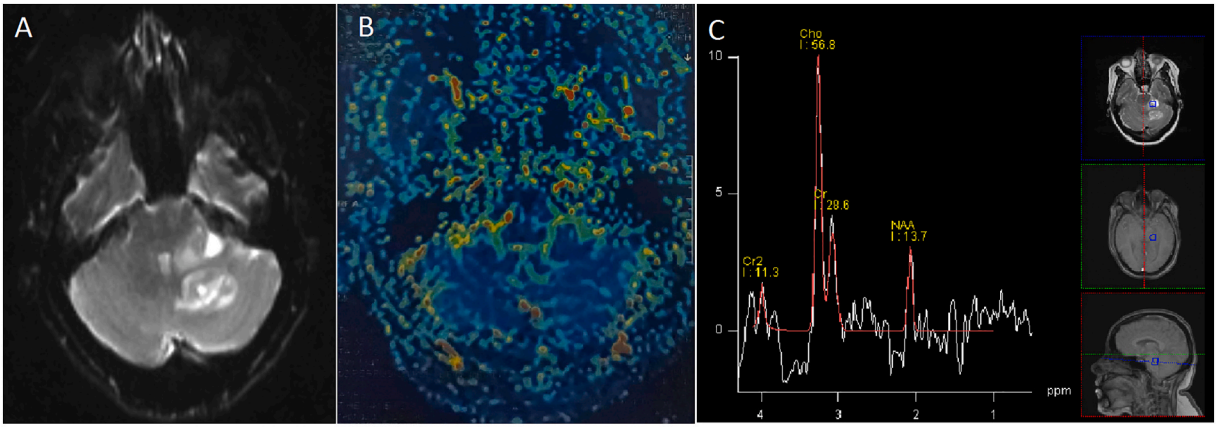

Fig. 2. Diffusion (A), Perfusion (B) and Spectroscopy (C) sequences of a brain MRI showing the multimodal parameters of the tumor.

metastases. An assessment of the tumor extent through a body CT scan was performed, revealing no other extra-cranial lesions.

The patient underwent surgery. She was placed on prone position. A medial suboccipital approach was performed, followed by a corticotomy on the superior pole of the left cerebellar hemisphere. This allowed an access to a greyish, friable, nonhemorrhagic, and nonencapsulated tumor. We went through a resection which was incomplete as the tumor was apparently infiltrating the cerebellar peduncle. Postoperative course was uneventful, and control CT scan (Fig. 3) showed no evidence for postoperative complications. The patient was discharged on the third postoperative day. Histopathologic exam (Fig. 4) found a tumoral proliferation with a high cellular density. It was composed of cells with moderate signs of anaplasia. Nuclei were oval, dense and hyperchromatic. Homer Wright rosettes with fibrillar center were present, as well as a microvascular proliferation. No signs of necrosis were shown, but adjuvant cerebellar tissue was infiltrated with a reactive gliosis. Immunohistochemistry showed a strong and diffuse positivity to Synaptophysine. Glial fibrillary acidic protein highlighted background reactive astrocytes, P53 was negative, and Ki67 was 60%. All these features concluded to a classic MB. The patient had a spine MRI that did not show any stigmata for metastatic dissemination. Adjuvant radiotherapy with a total dose of 54 Grays. A 6 months follow up did not show any signs of progression of the tumoral residue.

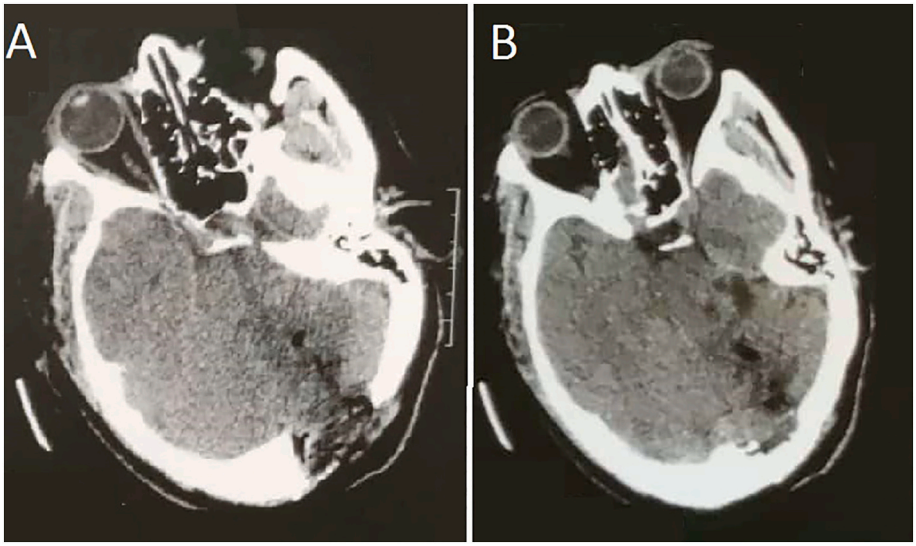

Fig. 3. Axial section of a postoperative brain CT scan without (A) and with (B) injection of contrast product showing the absence of any postoperative complication.

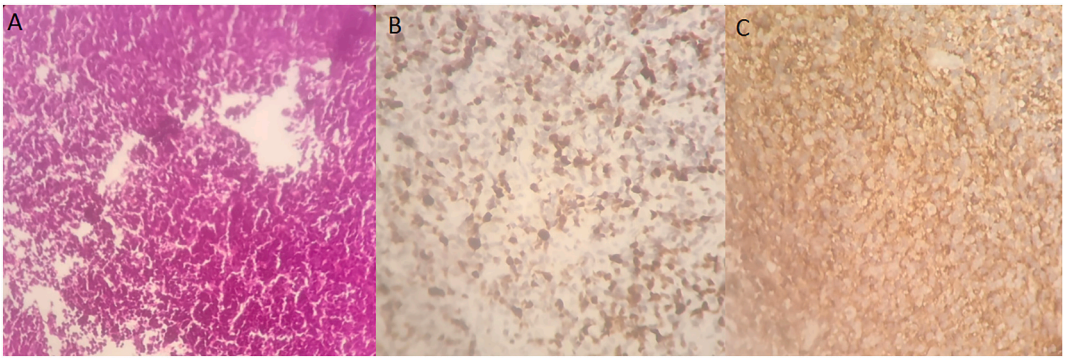

Fig. 4. Histological figures: microscopic overall study (A), Ki67 activity (B) and reaction to synaptophysine (C) concordant with the diagnosis of medulloblastoma.

Table 1
Previously published cases in the elderly (above the age of 60).

Author	Age/ sex	Location	Metastases	Treatment	Histologic diagnosis	Outcome
Seitz (1978) (9)	88/F	Vermis	Not precised	Surgery + radiotherapy	Classic	Alive after a 8 months follow up
Kepes (1987) (10)	73/F	Cerebellar hemisphere	No	Surgery + radiotherapy	Classic	Alive after a 4 months follow up
Cervoni (1994) (11)	71/M	Cerebellar hemisphere	No	Surgery + radiotherapy	Classic	Alive after a 42 months follow up
Cervoni (1994) (11)	67/M	Cerebellar hemisphere	No	Surgery + radiotherapy + chemotherapy	Classic	Alive after a 6 months follow up
Ramsay (1995) (12)	66/M	Vermis and cerebellar hemisphere	Yes	Surgery + radiotherapy	Classic	Dead after a 23 months follow up
Ramsay (1995) (12)	65/F	Cerebellar hemisphere	No	Surgery + radiotherapy	Desmoplastic	Alive after a 29 months follow up
Salvati (2000) (13)	68/M	Cerebellar hemisphere	No	Surgery + radiotherapy + chemotherapy	Classic	Alive after a 26 months follow up
Jaiswal (2000) (14)	65/M	Vermis	No	Surgery + radiotherapy + chemotherapy	Classic	Alive after a 3 months follow up
Yong (2006) (6)	71/M	Vermis	No	Surgery + radiotherapy	Classic	Alive after a 6 months follow up
Huppman (2009) (1)	66/M	Cerebellar hemisphere	No	Surgery	Classic	Not reported
Snuderl (2015) (7)	62/F	Cerebellar hemisphere	No	Surgery + radiotherapy	Classic	Not reported
Liang (2016) (3)	72/F	Cerebellar hemisphere	No	Surgery + radiotherapy	Classic	Not reported
De (2018) (4)	72/M	Not reported	No	Surgery + radiotherapy + chemotherapy	Classic	Alive after a 25 months follow up
Murase (2018) (2)	63/F	Vermis	No	Surgery + radiotherapy + chemotherapy	Classic	Alive after a 15 months follow up

3. Discussion

Most of the tumors of the posterior fossa are represented by metastases, which represent by far the most common intra-axial lesion at this age, followed by hemanglioblastomas and ependymomas [1,4]. MB are known to be pediatric tumors, as they represent 20 to 25% of the CNS malignancies in the childhood, and as 85% of the cases are diagnosed below the age of 15 [2,5]. MB may also be diagnosed in the young adult before the age of 30, with some radiologic and histological characteristics that are different from those reported in the children [1,2].

It is admitted by most of the published series that MB are extremely rare above the age of 40 [6]. MB diagnosed at the late elderly, after the age of 60 is exceptional, as we are only reporting the 15th diagnosed case. The previous 14 cases are resumed in Table 1.

Typical clinical presentation of MB is those of a quickly evolving mass of the cerebellum, traduced by signs of high intracranial pressure associated to symptoms of cerebellar dysfunction. The time of onset of the signs is classically counted in weeks. However, adult MB symptoms may progressively set up over months [7].

Generally, MB appears in imaging as a solid, strongly enhancing masses developing within medial structures of the vermis, with some tendency to infiltrate the 4th ventricle. In this sense, infiltration of the floor of the 4th ventricle is one of the major prognostic elements related to the MB. Adult MBs are more often located in the cerebellar hemisphere. This finding is supported by the previously reported cases, in which 10 of 14 lesions were located within the cerebellar hemisphere. Moreover, irregular contrast enhancement and the presence of cystic components within the tumor are frequently found in adults' MBs [2,6]. In the present case, the MRI showed two lesions located in the cerebellar hemisphere, with diffusion, perfusion and spectroscopic elements that suggested metastases at first sight. Based on these observations, we suggest that despite their rarity, and even if clinico-radiologic parameters evocative for other diagnoses, MBs should still be considered in the differential diagnosis of posterior fossa tumors in the adulthood.

Childhood MBs may be related to all of the four histological subtypes: classic, desmoplastic/nodular, extensive nodularity and large cell/anaplastic. Adulthood MBs are mainly represented by classic and desmoplastic subtypes [1,8]. This is confirmed by the previously reported cases, among which 13 were of a classic subtype, and one desmoplastic/nodular. Unfortunately, we do not dispose of a genetic analysis. Over the previously reported cases of MBs in the elderly, only two had a genetic analysis of the tumor, and both were non-WNT/non-SHH [2,5].

Therapeutic approaches for the MB in the childhood are well standardized. Depending on the age of the infant, chemotherapy and radiotherapy are proposed. Molecules of the chemotherapy and doses of the radiotherapy depend from risk stratification (quality of the resection, presence of stigmata of dissemination on CSF and on imaging, histological and genetic stratification) [9]. Concerning adulthood's MBs, therapeutic adjuvant approach is still controversial. In fact, the role of chemotherapy has been established in infants as it allows to reduce the radiation dose or to postpone radiotherapy when the child is aged below 3, its efficiency is still unclear in adults. In addition, adult patients are more vulnerable to chemotherapeutic toxicity, and side effects are more severe at this age [2,9]. Another fact is that, the chemotherapy is known to potentiate the effect of radiotherapy in children. This fact is still unclear in adults. This is why only an irradiation was propped for our patient. Previous experiences in terms of therapeutic protocols in elderly MBs were disparate. Thirteen over 14 patients had a radiotherapy, whilst a chemotherapy was added to irradiation in 5 cases.

4. Conclusions

Although only few cases of MBs in the elderly have previously been reported, this entity should remain in mind as clinical radiologic and histological features have different characteristics to those known in the childhood. Diagnostic and therapeutic tools identified in children are not always applicable in the elderly because tumors display similar and dissimilar features between groups of age. As population is aging worldwide, more reports of MB in the elderly are to emerge which may allow a better knowledge of the characteristics of this pathology at this age category, and better standardize therapeutic approaches.

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Patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethical approval

No ethical approval is needed for this publication.

Author contribution

GG wrote the manuscript; SG and MR made the bibliographic research; IZ and MB corrected the manuscript.

Research registration number

Not applicable.

Guarantor

The correspondent author is the guarantor for this manuscript.

Declaration of competing interest

The authors declare no conflict of interest.

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