Case report

DIFFUSION-WEIGHTED MR IMAGING AND PATHOLOGIC FINDINGS IN ADULT CEREBELLAR MEDULLOBLASTOMA

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SUMMARY

Objectives, materials and methods: The authors present the diffusion-weighted MR imaging and pathologic findings in two adult patients with cerebellar medulloblastoma.

Results: Both presented with a vermian mass of the posterior fossa with low signal on SE T1 weighted images, and moderate enhancement of the mass after gadolinium injection. The tumors were of high intensity on diffusion-weighted images with low ADC value. The ADC values ($\times 10^{-3}\text{mm}^2/\text{s}$) were respectively 0.60 $\pm$ 0.06 and 0.59 $\pm$ 0.11 (tumor), and 0.65 $\pm$ 0.04 and 0.67 $\pm$ 0.07 (cerebellar white matter). Tumors were highly cellular and composed of densely packed small round cells with hyperchromatic nuclei and scanty cytoplasm.

Conclusion: diffusion-weighted MR imaging may be useful for the diagnosis of cerebellar medulloblastoma, due to their high cellularity and high nuclear-to-cytoplasmic ratio.

Key words: diffusion MRI, medulloblastoma, PNET, adults.

RÉSUMÉ

Médulloblastomes cérébelleux de l’adulte et IRM de diffusion

Objectif et matériel et méthodes : Les auteurs présentent deux cas de médulloblastome cérébelleux de l’adulte explorés par IRM avec séquence de diffusion.

Résultats : Les deux patients présentaient une lésion vermienne visible en hyposignal T1, modérément rehaussée après injection de Gadolinium. Les tumeurs présentaient un hypersignal en diffusion avec un coefficient apparent de diffusion diminué. La mesure du coefficient apparent de diffusion était respectivement 0,60 $\pm$ 0,06 et 0,59 $\pm$ 0,11 (tumeur), et 0,65 $\pm$ 0,04 et 0,67 $\pm$ 0,07 (substance blanche du cervelet). L’examen histopathologique montrait une prolifération tumorale dense composée de petites cellules avec un cytoplasme peu abondant.

Conclusion : Les séquences de diffusion en IRM peuvent être utiles pour le diagnostic de médulloblastome chez l’adulte en raison d’une prolifération tumorale dense et d’un rapport nucléo-cytoplasmique élevé.

Mots-clés : Imagerie de diffusion, médulloblastome, tumeur neuroépithéliale primitive, adultes.

INTRODUCTION

Medulloblastoma is the most frequent neoplasm of the posterior fossa in childhood, while it is rare in adult patients [3]. The vermis is the most frequent location [3]. Medulloblastomas are composed of undifferentiated neuroectodermal cells with high cellularity [3]. These tumors are classified as highly malignant due to their locally aggressive behaviour and subarachnoid dissemination. Diffusion-weighted (DW) imaging has been used to study intra-axial tumors (gliomas, lymphomas) and brain abscesses [6, 10]. High signal with diffusion-weighted MR imaging has only been reported in cases of cerebellar medulloblastoma in children [7, 8, 13]. In this report, we describe DW MR and pathologic findings in two adult patients with cerebellar medulloblastoma.

CASE REPORTS

Patient 1

A 42-year-old woman was referred for an MRI examination of the brain because of progressive headache increasing over two months.

CT showed a midline mass of the posterior fossa. The lesion was spontaneously hyperdense with mild enhancement after intravenous contrast administration.

MRI (1.5Tesla) (figure 1) showed a midline lesion with low signal on SE T1-weighted images, and moderate high signal on PD and TSE T2-weighted images. The tumor was isointense on FLAIR images. No MRI finding suggestive of hemorrhage was observed within the mass on SE T1-weighted and GE T2-weighted images. After gadolinium injection, enhancement of the mass was moderate and homogeneous, and a satellite nodule was clearly visible. The diffusion weighted images (DWI) were acquired with an echo-planar imaging sequence (b values $= 0$ and 1000). We recorded the ADC values from the solid portion of the tumor, from peritumoral hyperintense areas on FLAIR images, and from the normal cerebellum. The tumor was of high intensity on DWI. The ADC values ($\times 10^{-3}\text{mm}^2/\text{s}$) were 0.60$\pm$0.06 (tumor), 0.65$\pm$0.04 (cerebellar white matter), and 1.13$\pm$0.19 (surrounding edema).

The patient underwent a suboccipital craniotomy, and complete removal of the tumor was achieved via a trans-vermian route. The tumor had
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a well-demarcated grey homogeneous appearance. On microscopic examination, the lesion was not well circumscribed. It was highly cellular and composed of densely packed small round cells with hyperchromatic nuclei and scanty cytoplasm. Neuroblastic rosettes were absent. Mitotic activity was marked (up to 15 mitoses per 10 high power fields). Thin fibrous vascular septa without edema were inter-
spersed within the tumor. Immunohistochemically, some tumoral cells showed cytoplasmic reactivity for GFAP (clone 6F-2, Dako, 1/10) and synaptophysin (clone Klon, Dako, 1/10), but were negative for neurofilament proteins (clone 2F11, Dako, 1/25), cytokeratins (clone KL1, Immunotech, 1/100), and CD45 (clone PD7/26 and 2B11, Dako, 1/50). These histologic findings corresponded to those of a classic medulloblastoma. After the surgery, the patient’s neurologic symptoms and signs gradually improved.

Patient 2

A 22-year-old woman presented with intense headaches increasing over one month, vomiting and ataxia. MRI (1.5Tesla) (figure 2) showed a vermian lesion with low signal on SE T1-weighted images, moderate high signal on PD, TSE T2 and FLAIR images with little surrounding edema. Compression of the fourth ventricle was resulted in non communicating hydrocephalus. No MRI finding suggestive of hemorrhage was observed within the mass on SE T1-weighted and GE T2-weighted images. After gadolinium injection, enhancement of the mass was moderate and homogeneous. The tumor was of high intensity on DWI. The apparent diffusion coefficient (ADC) values ($\times 10^{-3}$mm$^2$/s) were 0.59±0.11 (tumor), 0.67±0.07 (cerebellum white matter), and 1.10±0.04 (surrounding edema).

The patient underwent a suboccipital craniotomy, and partial removal of the tumor was achieved via a trans-vermian route. Due to the proximity of the torcular, the residual fragment was coagulated. The tumor had a well-demarcated grey homogeneous appearance. On microscopic examination, the lesion was composed of densely packed small round cells with hyperchromatic nuclei and scanty cytoplasm. Neuroblastic rosettes were present. Mitotic activity was marked (up to 15 mitoses per 10 high power fields). Immunohistochemically, the findings were similar to those described for the first case and corresponded to a classic medulloblastoma. After the surgery, the patient’s neurologic symptoms and signs improved.

DISCUSSION

Medulloblastoma is a small cell primitive neuroectodermal tumor (PNET) of the cerebellum [3]. Medulloblastoma is the most frequent malignant central nervous system neoplasm within the first 15 years of life [3]. The cerebellar vermis is by far the most frequent location. The MR findings of medulloblastoma in children are dominated by an increased relaxation time on T1- and T2-weighted sequences [12]. In adults, medulloblastoma is rare and the cerebellar hemispheres are its most frequent location. The tumors are typically hypointense on T1-weighted images but a spectrum has been described on T2-weighted images [1, 5]. Although there is no pathognomonic MR appearance of adult cerebellar medulloblastoma, the finding of a well-demarcated, mildly to moderately enhancing hemispheric mass involving the cerebellar surface in a young adult is suggestive of medulloblastoma [5]. The frequent occurrence of isointense T2 signal to the grey matter has been attributed to the marked cellularity, high nuclear-to-cytoplasmic ratio, intratumoral hemorrhage or occasionally the desmoplastic fibrocollagenous response elicited by dural invasion [9]. Differential diagnosis should include other...
markedly hypercellular neoplasms with relatively low intensity on T2-weighted images: other undifferentiated round cell tumors (pinealblastomas and neuroblastomas), and other tumor types, including most lymphomas, mucinous adenocarcinomas and amelanotic melanoma metastases [9].

In our cases, both tumors showed high signal intensity on diffusion weighted images with slightly restricted ADC. These findings were suggestive of high cellularity [4]. The ADC values were 0.59 and 0.60 (×10⁻³mm²/s), in the range of values described in lymphomas [10] [4] and meningiomas [6], but lower to the values described in association with grade II and high grade astrocytomas and glioblastomas [4, 6]. Well-circumscribed brain lesions with a decreased apparent diffusion coefficient and a slightly or moderately increased signal on T2-weighted images have also been found in patients with metastases from a small-cell bronchial carcinoma, from a pulmonary adenocarcinoma, or from a breast carcinoma [2].

The ADC values in peritumoral hyperintense areas on T2WI were 1.13 and 1.10 (×10⁻³mm²/s), and were much higher than the ADC values into the tumor. These peritumoral areas corresponded probably to edema rather than true neoplastic cell infiltration according to Tien et al [11]. However, the ADC values in peritumoral hyperintense areas were in the range of values observed in glioblastoma and meningioma [6]. Kono et al’s findings seem disheartening because they do not support the hypothesis that peritumoral cell infiltration can be depicted by ADCs or ADC map.

Our cases demonstrate the utility of diffusion weighted imaging in the diagnosis of cerebellar medulloblastomas in adults due to their high cellularity and high nuclear-to-cytoplasmic ratio [4]. Nevertheless, from a histopathological point of view, since medulloblastoma presents morphologically as an undifferentiated round-cell tumor, the main differential diagnosis in an adult should include metastatic small cell carcinoma, especially of the lung, and malignant lymphoma. The morphological pattern of these tumors is often very close. Neuroblastic rosettes are helpful to distinguish medulloblastoma, but they are inconistent. Therefore, immunohistochemical study is helpful in order to distinguish these tumors from medulloblastoma. Indeed, keratin staining is positive in small cell carcinoma contrary to medulloblastoma and CD45 staining is positive in lymphoma while it is negative in medulloblastoma. Glioblastoma with poorly differentiated round cells is the other diagnosis that has to be ruled out. As opposed to medulloblastoma, more differentiated and typical areas of glioblastoma are usually discernible, at least focally. GFAP-immunoreactivity is not distinctive [3].

In conclusion, differential diagnosis of adult posterior fossa tumors with high signal on diffusion weighted images should include medulloblastomas. Their appearance is not pathognomonic, and may be similar to lymphomas, metastases (small cell carcinomas), and glioblastomas with poorly differentiated round cells. Low ADC values indicate high cellularity and high nuclear-to-cytoplasmatic ratio, which might be helpful in order to distinguish medulloblastomas from glioblastomas.

REFERENCES