Child's Nervous System (2019) 35:871–874
https://doi.org/10.1007/s00381-019-04102-6

CASE REPORT

Multimodel MRI features of an intracranial juvenile Xanthogranuloma

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Received: 29 December 2018 / Accepted: 20 February 2019 / Published online: 28 February 2019

Abstract

Juvenile xanthogranuloma (JXG) is a benign, self-limiting histiocytic disorder of infancy and early childhood, usually presented as a single or multiple cutaneous lesions. The central nervous system is rarely affected by JXG. There were only a few reports of intracranial JXG cases which described its features on MR spectroscopy (MRS) and diffusion-weighted imaging (DWI), but its features on susceptibility-weighted imaging (SWI) and perfusion-weighted imaging (PWI) have not been reported yet. Here, we reported an intracranial JXG case which underwent multimodal MRI examinations including DWI, SWI, and PWI. The multimodal MRI provided a thorough insight into this disease and we found that intense enhancement and high perfusion may be important clues for the diagnosis.

Keywords Xanthogranuloma, juvenile · Magnetic resonance imaging · Perfusion-weighted imaging · Diffusion-weighted imaging · Susceptibility-weighted imaging

Introduction

Juvenile xanthogranuloma (JXG) is one of the most common forms of non-Langerhans cell histiocytosis in young children. This proliferative disorder is typically noted in the first two decades of life and is primarily characterized by lesions in the skin or soft tissue in the majority of patients [1]. The typical clinical course of JXG is self-limiting and uncomplicated, but systemic JXG—especially JXG in the central nervous system (CNS)—is generally difficult to treat and is often fatal [2]. Pathological examination remains the gold standard for the diagnosis of intracranial JXG. Some immunohistochemical markers like CD68 (clone: PGM1) and CD163 represented the markers of histiocytic differentiation. The absence of S100 and/or CD1a immunostaining excluded the diagnosis of Langerhans cell histiocytosis (LCH) [2]. Only several intracranial JXG cases were reported in literature whose features on MR spectroscopy (MRS) and diffusion-weighted imaging (DWI) were discussed. This is the first report of susceptibility-weighted imaging (SWI) and perfusion-weighted imaging (PWI) features of an intracranial JXG.

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Case report

A 7-year-old girl was admitted with dizziness, vomiting, and weakness of the right limbs, and without headache or convulsion of the limbs. A CT scan in the local hospital revealed a mass in the right cerebellum. Then she was referred to the university hospital. Routine laboratory examination failed to detect obvious abnormality. A thorough physical examination found inaccurate responses in the right side finger-nose test, but failed to reveal any cutaneous lesions or deformities.

MRI of the brain revealed a well-defined patchy area in the right cerebellum, which was mild hypointense on T1-weighted images and heterogeneously hyperintense on T2-weighted images, and mildly hyperintense on T2-flair images. It demonstrated intense and homogeneous enhancement after administration of Gadolinium (Fig. 1). No perilesional edema was noted. No abnormalities such as calcification, bleeding, or vascular malformation were revealed by SWI (Fig. 2a, b). Diffusion-weighted imaging (DWI) did not demonstrate restricted diffusion in the lesion (Fig. 2c, d). PWI demonstrated a high regional cerebral blood flow (rCBF), high regional cerebral blood volume (rCBV), delayed mean transit time

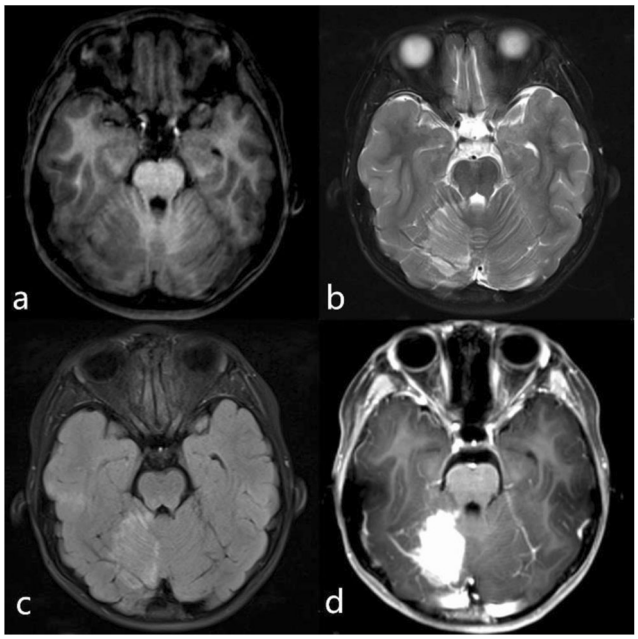

Fig. 1 Conventional MRI of intracranial juvenile xanthogranuloma. The lesion was mildly hypointense on T1WI (**a**), heterogeneously hyperintense on T2WI (**b**), and mildly hyperintense on T2-flair images (**c**). Contrast-enhanced T1WI shows intense enhancement of the lesion (**d**)

(MTT), and decreased time to peak (TTP) compared to the contralateral cerebellum (Fig. 3).

The patient underwent craniotomy and the lesion was found to be located in the right cerebellar hemisphere. It was

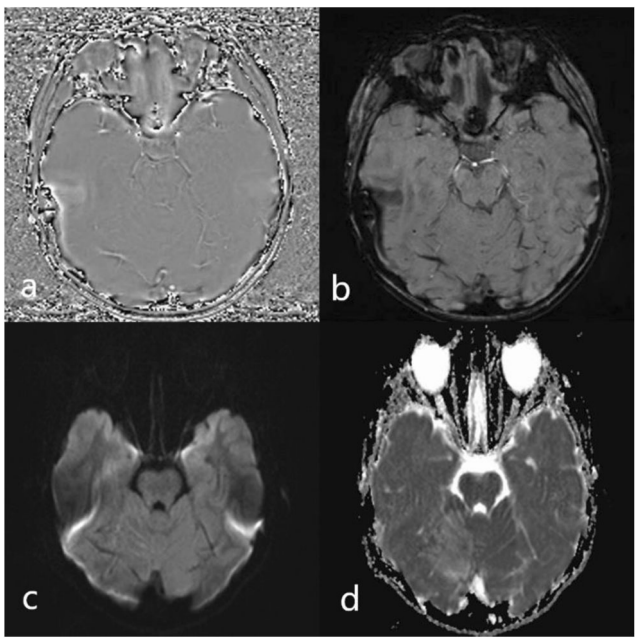

Fig. 2 Susceptibility and diffusion weighted imaging (SWI and DWI) of intracranial juvenile xanthogranuloma. Phase (**a**) and SWI (**b**) images does not reveal obvious abnormalities within the lesion. It was isointense on DWI (**c**, **b** = 1000) and slightly hyperintense on apparent diffusion coefficient (ADC) map (**d**)

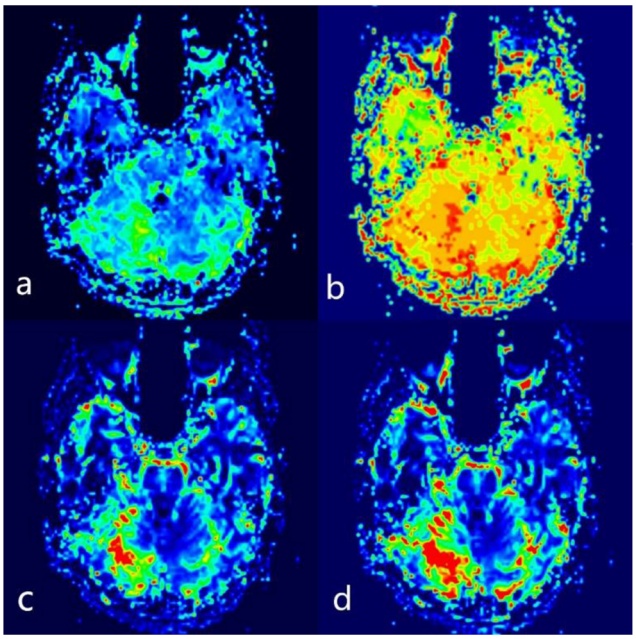

Fig. 3 Perfusion-weighted imaging of intracranial juvenile xanthogranuloma. MR perfusion showed hyperperfusion, with a maximum relative cerebral blood flow (rCBF) value of 3.2, a maximum relative cerebral blood volume (rCBV) value of 5.7, a relative mean transit time (rMTT) value of 1.65, and a relative time to peak (rTTP) value of 1.14

brittle and waxy, and its color was dark and red. A part of the tumor tissue was clamped for pathological examination, and the lesion was completely removed.

Microscopic examination showed proliferation of histiocytes and a few multinucleated giant cells. Some angiogenesis was also observed. Immunohistochemical staining was positive for CD68-PGM1 and CD163, and the ki-67-labeled proliferation index was 20–25%. Staining for S100 was negative. The marker of CD1a, which is an important marker to differentiate JXG from LCH, was negative [3]. Genetic testing did not find BRAF mutation. These findings suggested the diagnosis of JXG (Fig. 4).

Discussion

MRI can display the morphological characteristics of lesions, and reflect the functional changes as well, including the blood perfusion, cellularity, susceptibility, and metabolism. Multimodal, functional MRI plays an increasingly important role in the diagnosis of cerebral infarction, neural degenerative diseases and tumors. Some similarities together with differences were noted when we compared the MRI features of the present case with previously reported cases [2, 4].

JXG was reported to involve the ventricles, sellar region, cerebellopontine angle, intra-parenchyma, dural-cortical interface, and skull base. Solid lesions are more common. It was usually isointense on T1WI and T2WI, and was

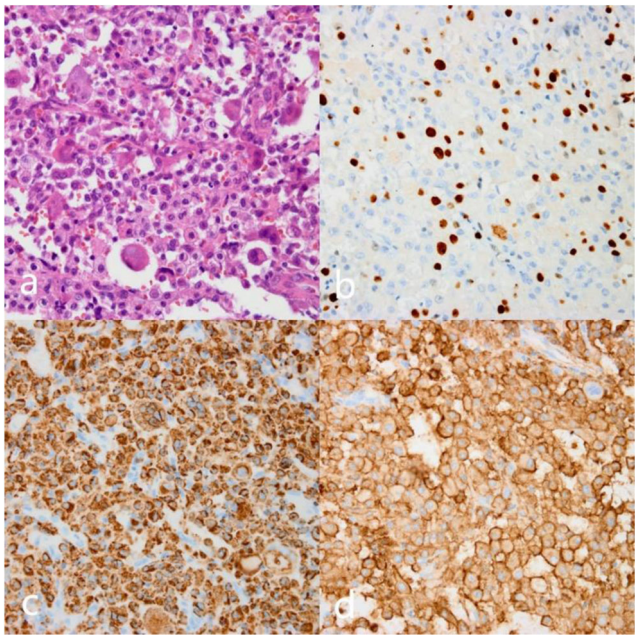

Fig. 4 Microscopic examination of intracranial juvenile xanthogranuloma. Hematoxylin-eosin staining (× 40) shows proliferation of histiocytes (**a**). The ki-67 labeled proliferation index was 20–50% (**b**). CD 68 immunostaining was positive (**c**). CD 163 immunostaining was positive (**d**)

homogeneously enhanced after administration of Gadolinium [2]. In the current case, the lesion was located at the cerebellar hemisphere and showed mildly hypointense on T1WI and heterogeneously hyperintense on T2WI. These features are different from the previous reports. The lesion showed homogeneous and intense enhancement with Gadolinium which was consistent with the literature. Our case was more challenging to diagnose than previous cases because it was a patch-like lesion without significant mass effect, which has not been seen in the literature.

Diffusion and perfusion characteristics have been used to differentiate various brain tumors. Some previous JXG cases underwent DWI and they demonstrated hyperintensity [3, 5–7]. In this case, however, the lesion was isointense on DWI and slightly hyperintense on ADC map. The perfusion characteristics of JXGs have not been reported. In this case, PWI showed an increase of rCBV, rCBF, MTT, and TTP in the lesion compared to the contralateral normal side. In view of the microscopic examination, the increased perfusion may be related to the angiogenesis within the tumor. SWI features also have not been reported in the literature. In our patient, we failed to find any obvious abnormalities on SWI.

The first differential diagnosis was the cerebellar infarction because this path-like lesion did produce significant mass effect. Infarction can also demonstrate hypointensity on T1WI and heterogeneous hyperintensity on T2WI, with intense enhancement. The increased perfusion was helpful to exclude cerebellar infarction since infarction usually demonstrates decreased rCBV and especially rCBF. In addition, restricted diffusion is often observed in infarction which provided a clue to differentiate it from our case.

This case had been considered as dysplastic gangliocytoma of the cerebellum (Lhermitte-Duclos disease, LDD) before surgery. The MRI imaging feature of LDD is the characteristic appearance of alternating hypointensity and hyperintense stripes (tiger striped). The lesion of this case demonstrated slightly high signal intensity with striated appearance on T2WI, which was somewhat like "tiger striped." And LDD could also demonstrate elevated rCBV [6] just like this case. However, LDD usually presented as a non-enhancing lesion which was not consistent with the current case.

Intracranial JXG is a rare disease, so little evidence exists to guide the management. Surgery was reported to be a recommendable treatment in patients with accessible solitary lesions. Patients with surgically inaccessible or multi-centric lesions can be treated with corticosteroids, chemotherapy, radiation, or a combination treatment. More studies are needed to assess the efficacy of these treatments [8].

Conclusion

This report demonstrates the imaging features of intracranial JXG and enriches our knowledge with the multimodal, functional MRI findings. Intense enhancement and high perfusion may be important clues for the diagnosis although these changes do not seem to be characteristic. More cases with multimodal MRI are needed to replicate our findings.

Compliance with ethical standards

Conflict of interest We declare that we do not have any commercial or associative interest that represents a conflict of interest in connection with the work submitted.

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