Case Report

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Journal of Clinical Radiology and Case Reports

2018
Vol.2 No.1:01

Spontaneous Remission of Vogt-Koyanagi-Harada Meningoencephalitis

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Rec date: January 10, 2018; **Acc date:** January 16, 2018; **Pub date:** January 24, 2018

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Citation: Abdullayev N, Maus V, Neuhaus V, Christoph K, Borggrefe J, et al. Spontaneous remission of Vogt-Koyanagi-Harada Meningoencephalitis. J Clin Radiol Case Rep 2018, Vol.2 No.1:04.

Abstract

We describe a 65-year-old patient with Vogt-Koyanagi-Harada (VKH) syndrome and primary malignant melanoma of the trunk presenting with systemic tumor progress and new contrast-enhancing intracranial lesions without neurological symptoms. While the systemic melanoma disease ultimately was in progress, the correct radiological interpretation of brain lesions as intracranial manifestation of VKH was mandatory to prevent the patient from unnecessary whole brain radiotherapy.

Keywords: Vogt-koyanagi-harada; Meningoencephalitis; Cerebral manifestations of VKH; Vitiligo

Case Report

A 65-year-old patient with a 10-year history of VKH disease including vitiligo and uveitis and a 27-year history of primary malignant melanoma of the trunk exhibited histologically proven mediastinal melanoma masses (BRAF V600E-Mutation, American Joint Committee on Cancer stage IV) during follow-up. Initial brain MRI was normal. Treatment with BRAF and MEK inhibitors was initiated with partial response in the 2-months chest and abdominal CT control-scan. Brain MRI control-scan revealed new leptomeningeal contrast enhancement of the pons and T2 hyperintense lesions in the right cerebral peduncle without neurological symptoms **(Figure 1)**.

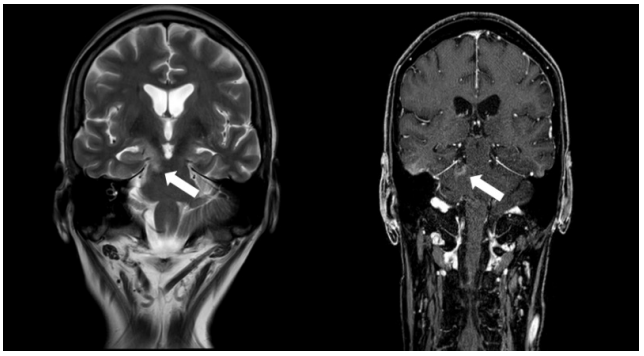

Figure 1 Leptomeningeal contrast enhancement of the pons and T2 hyperintense lesions in the right cerebral peduncle (left-T2 TSE coronar, right-T1 3D FFE SPIR coronar).

Treatment of melanoma remained unchanged and further control-MRI of the brain after 2 months showed absence of contrast enhancement with regressive T2 lesions. New contrast-enhancing lesions were present in the supratentorial white matter **(Figure 2)**.

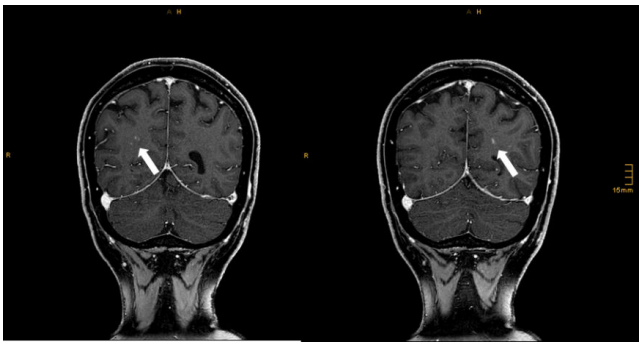

Figure 2 Contrast-enhancing lesions in the supratentorial white matter (T1 3D FFE SPIR coronar).

Mediastinal melanoma masses showed remission on CT. Morphological findings and clinical course were deemed atypical for brain melanoma metastasis and accepted as cerebral manifestation of VKH syndrome. Probatory steroid therapy was recommended, but rejected by the patient. Nevertheless, a further 3-months control-MRI of the brain demonstrated complete remission of all contrast-enhancing lesions and slight residual T2 hyperintensity in the cerebral peduncle **(Figure 3)**. Melanoma metastases were dramatically progressive on CT during unchanged therapeutic regimen.

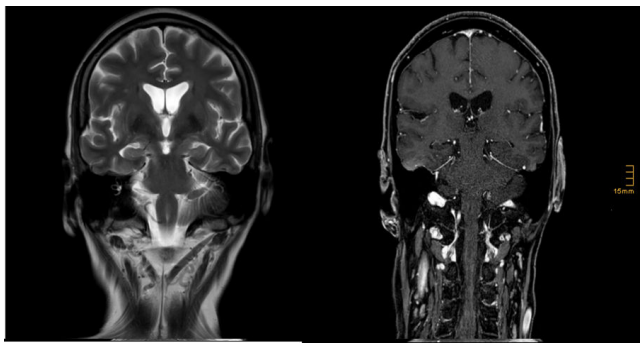

Figure 3 Residual T2 hyperintensity in the right cerebral peduncle and remission of leptomeningeal contrast enhancement of the pons (left-T2 TSE coronar, right-T1 3D FFE SPI.

Discussion

VKH syndrome is a granulomatous inflammatory disorder mainly affecting tissues containing pigmented cells (ocular, auditory, integumentary). Patients often present with vitiligo, alopecia, auditory signs, and meningeal irritation [1]. Th1 and Th17 $CD4^+$ lymphocytes seem to play a pivotal role in the pathogenesis of VKH disease [2].

The natural history can be divided into 4 stages: prodromal, acute-uveitic, chronic, and chronic recurrent and is characterized by bilateral uveitis with multifocal retinal detachment and iridocyclitis. The involvement of the central nervous system is rare and may be associated with focal neurological signs [3].

Neuroimaging (MRI) is mandatory to confirm intracranial manifestation of VKH, although abnormal MRI findings have been reported in a few cases only. Early reports described diffuse leptomeningeal contrast enhancement and T2-bright periventricular lesions, while more recent ones described lesions within the brain stem and peduncle as well as pachymeningeal enhancement [4,5].

We here present a patient with progressive malignant melanoma and VKH syndrome exhibiting multiple new contrast-enhancing cerebral lesions and leptomeningeal enhancement with spontaneous remission. Interestingly, our patient showed a very late onset of leptomeningeal involvement without neurological symptoms, although abnormal signal changes and pathologic contrast-enhancement were located in the right cerebral peduncle. Hashimoto et al. described similar changes mainly in the posterior fossa as preferred localization [6].

In the present case, the opposed systemic course of the progressive melanoma disease under continuous immunotherapy in contrast to the initially new-onset intracranial lesions, that finally resolved spontaneously, led to the correct interpretation of these findings. In conclusion, cerebral manifestations of VKH in patients with primary malignant melanoma have to be delimited from melanoma metastasis as misinterpretation of clinical and MRI findings may lead to erroneous treatment, e.g., whole brain radiotherapy.

Conflict of Interest

The authors declare no conflict of interests.

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