

The diagnosis of Marburg Disease is course-dependent

Abstract

Marburg Disease, the fulminant form of multiple sclerosis, is a rare disease that typically kills within a year. We had a 38-year-old African American male who presented with right footdrop and was pathologically diagnosed with Marburg Disease. The patient recovered clinically after surgery and stayed stable for more than a year. The diagnosis of Marburg Disease was thus degraded.

Keywords: Marburg Disease, pathological diagnosis, clinical course

Zusammenfassung

Die akute Variante (Marburg) der multiplen Sklerose ist eine fulminant verlaufende, seltene Erkrankung, die gewöhnlich innerhalb eines Jahres zum Tode führt. Uns stellte sich ein 38 Jahre alter männlicher Afro-Amerikaner mit einer Fußheberparese vor. Es wurde eine Marburg-Variante der multiplen Sklerose pathologisch diagnostiziert. Nach dem Eingriff erholte sich der Patient klinisch und sein Zustand blieb für mehr als ein Jahr stabil. Die Diagnose Marburg-Variante der multiplen Sklerose wurde folglich herabgestuft.

Schlüsselwörter: Marburg-Variante der multiplen Sklerose, pathologische Diagnose, klinischer Verlauf

Background

Marburg Disease is an acute form of multiple sclerosis (MS) that is marked by rapid onset and often continual progression of the demyelination process. The National Multiple Sclerosis Society estimates the prevalence of MS to be around 135 per 100,000 population in the U.S., and a total MS population of approximately 400,000 [1]. Fortunately, Marburg Disease is very rare and may be encountered once in a neurosurgeon's career.

Case presentation

Our patient is a 38-year-old African American male who was admitted because of right footdrop. His past medical history was significant for hypertension, diabetes mellitus, and rheumatoid arthritis. He was on no medications at home. He was given a medication for rheumatoid arthritis back in the early 2000s but discontinued it shortly after because of side effects. He could not remember what it was. On physical exam, strength was 5/5 in the upper extremities and left lower extremity and 2/5 in the right lower extremity. Deep tendon reflexes were 1–2 and symmetric. Sensation to light touch and pinprick was intact.

On MRI of the brain, there was a 2.9 x 2.5 x 2.3 cm cystic mass in the left superior medial frontal lobe. This was hypointense on T1 and hyperintense on T2 with surrounding edema. After contrast administration, there was minimal peripheral enhancement (Figure 1). The differential

diagnosis included brain tumor and brain abscess. The patient denied any headaches despite the brain findings. The patient was given intravenous dexamethasone and a seizure medication. Metastatic workup was ordered and medical consultation was requested for possible infectious etiology. The patient underwent bone scan and CT of the chest, abdomen, and pelvis. They were all negative. His erythrocyte sedimentation rate was 1 mm/hour and white blood count $6.64 \times 10^9/L$. The patient was started on metronidazole and ceftriaxone. He eventually underwent craniotomy with decompression of the cystic lesion. Frozen section came favoring infectious etiology over glioma. After surgery, the patient was weaned from dexamethasone but continued on intravenous antibiotics. The final pathology report came with the diagnosis "acute demyelinating disease, known as Marburg's disease or Marburg's variant of multiple sclerosis." Hematoxylin and eosin sections revealed a process exclusively involving the white matter with sharp demarcation between the gray and white matter. The white matter appeared mostly replaced by histiocytes and reactive astrocytes (Figure 2A). There was an accompanying perivascular lymphocytic infiltrate (Figure 2A). CD68 immunohistochemistry demonstrated the histiocytes exclusively in the white matter with sparing of the gray matter (Figure 2B). The differential diagnosis would have been subacute ischemic process, however, neurofilament immunohistochemical tests revealed preserved axons within the lesion consistent with a demyelinating disease. Grocott's Methenamine Silver (GMS) special stain and culture were negative for fungal infection.

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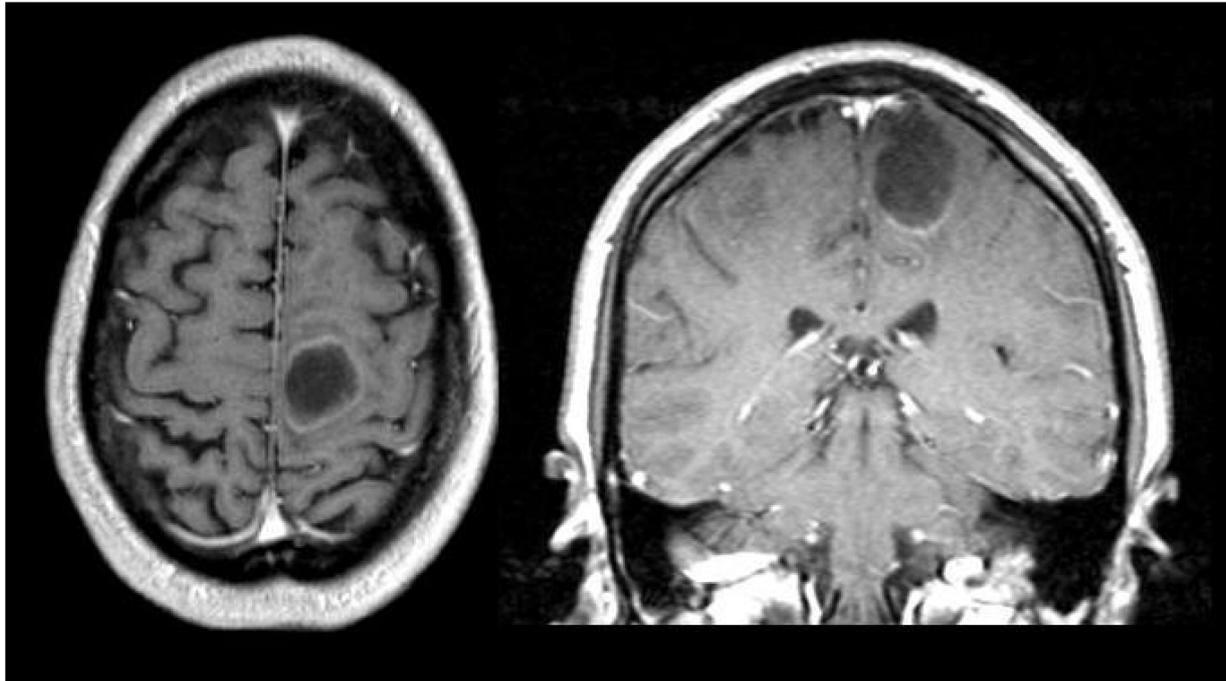


Figure 1: MRI of the brain showing a 2.9 x 2.5 x 2.3 cm hypointense mass in the left superior medial frontal lobe with minimal peripheral enhancement

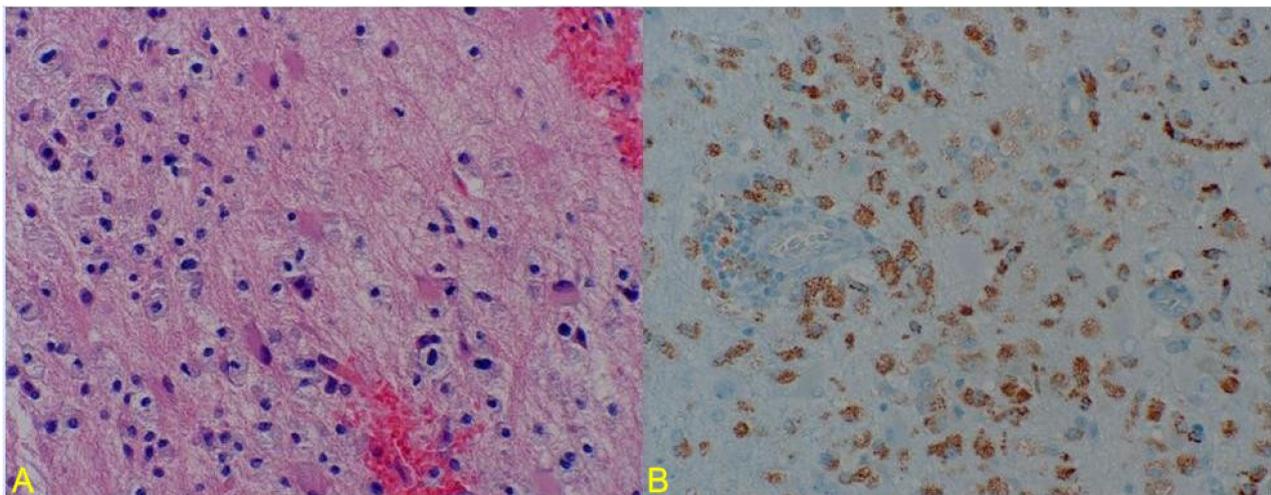


Figure 2: Pathologic view

A: Hematoxylin & eosin staining showing white matter with a macrophagic infiltrate and reactive astrocytes.

B: CD68 immunohistochemical staining showing positivity in the histiocytes which are exclusively in the white matter with sparing of the gray matter.

The patient was discharged home and a year and a half later is happily alive and doing well.

Discussion

Marburg Disease, an acute, fulminant, monophasic variant of multiple sclerosis, typically kills within a year [2], [3]. Demyelination usually begins in the hemispheres or the brainstem. Distribution of the lesions is usually indistinguishable from classical multiple sclerosis, but unlike classical MS, they occur simultaneously in all affected areas. On histological examination, lesions are generally

similar to those of classical MS though they tend to have more inflammatory infiltrates and appear more destructive [4]. If Marburg Disease occurs in the form of a sole big lesion it can be radiologically indistinguishable from a brain tumor or abscess. In our case, there was no fever, the erythrocyte sedimentation rate and white blood count were not elevated at admission, and culture of the biopsied material was negative. The patient was withdrawn from antibiotics when the final pathology report came confirming a demyelinating disease.

It is known that multiple sclerosis is more common in whites; however, when the disease happens in African Americans it is usually more severe [5]. Our patient is an

African American. Nobody yet knows for sure why these racial differences exist but many researchers think it is due to differences in the immune system. The rheumatoid arthritis in our patient's history is a strong indicator of an autoimmune process going on in the patient's organism. Although the pathological picture in our case was consistent with the Marburg variant of multiple sclerosis, the diagnosis would depend on the progression of the disease. Our patient have survived the critical stage and seems to be doing fine. The diagnosis will be downgraded to multiple sclerosis - chronic form if the patient follows a remitting course.

Three learning points can be extracted from this case:

1. The Marburg variant of multiple sclerosis is rare and requires high index of suspicion.
2. The Marburg variant of multiple sclerosis should be included in the differential diagnosis of abscess- and tumor-like lesions on MRI.
3. The diagnosis of the Marburg variant of multiple sclerosis should be made based on disease course.

Notes

Conflicts of interest

None declared.

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Please cite as

Walid MS, Sanoufa M. The diagnosis of Marburg Disease is course-dependent. *GMS Ger Med Sci.* 2010;8:Doc06.
DOI: 10.3205/000095, URN: urn:nbn:de:0183-0000958

This article is freely available from

<http://www.egms.de/en/gms/2010-8/000095.shtml>

Received: 2009-10-23

Published: 2010-03-02

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