

## Fulminant Multiple Sclerosis, What To Do? Diagnostic Approach and Management Options

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**Background:** Fulminant multiple sclerosis (MS) is the most aggressive type of MS, which typically leads to great disability and even death within weeks. It is usually monophasic and represents a medical emergency that calls for urgent management. Fulminant MS includes two variants, Marburg type and Balò concentric sclerosis. The Marburg type, which is also called tumefactive MS, is characterized by tumor-like demyelinating brain lesions while the Balò subtype is characterized by concentric layers of alternate myelination and demyelination.

**Methods:** A systematic literature review has been done to identify the cases of fulminant MS. Our objectives were to evaluate the diagnostic strategies and therapeutic approach and to highlight the characteristic clinical, pathological, and radiological features of fulminant MS.

**Results:** (1) Pathological features: Balò's variant was characterized by alternate concentric layers of demyelination and preserved myelin. While the Marburg subtype was characterized by demyelination associated with profound inflammation, tissue destruction, necrosis, and macrophage infiltration. (2) Presentation: fulminant MS is more common in young and usually monophasic. The symptoms may start with fever in some cases followed by a decreased level of consciousness and focal neurological deficit. Seizure has also been reported. Peripheral nervous involvement has also been reported in some cases of Marburg subtype. (3) Magnetic resonance imaging (MRI) with contrast: the Marburg variant is characterized by large lesions of tumefactive demyelination with mass effect and incomplete ring enhancement, but rarely presented with disseminated small demyelinating lesion. Remyelination of affected areas is rare. While in Balò concentric sclerosis, round lesions with irregular concentric rings of low and high signal alternate to give the onion bulb appearance. Peripheral enhancement indicates active demyelination. It may also be associated with peripheral diffusion restriction.

**Conclusion:** Fulminant MS is a rare type of MS which has a grave prognosis especially if not diagnosed and treated promptly and appropriately. Other fulminant demyelinating disorders as well as viral encephalitis, progressive multifocal leukoencephalopathy (PML), high-grade gliomas, and lymphomas need to be excluded.