

The component features of OMS include repeated, random and rapid eye movements in both horizontal, vertical and diagonal directions (opsoclonus); unsteady gait or loss of ability to stand and walk (ataxia); brief, repeated, shock-like spasms of several muscles within the arms, legs (myoclonus), or tremor interfering with hand use. Behavioral and sleep disturbances, including extreme irritability, inconsolable crying, reduced and fragmented sleep (insomnia) and rage attacks are common. Difficulty articulating speech (dysarthria), sometimes with complete loss of speech and language may occur. Additional symptoms such as decreased muscle tone (hypotonia) and vomiting are common. The most common cause of OMS in young children is paraneoplastic. A small, often hidden tumor presumably provokes the immune system into attacking the nervous system, which may also control the tumor or even cause it to regress. Tumors are NOT in the brain, but are in other areas of the body, usually in chest or abdomen. In 50-80 percent of affected young children, a tumor of embryonic nerve cells (neuroblastoma or ganglioneuroblastoma) is responsible for the symptoms associated with OMS. In other affected individuals, the disorder has been designated 'idiopathic' or attributed to various mostly viral infections. However, the high rate of spontaneous tumor regression means that the tumor may be gone before it is looked for. In older children or teens, viral infections are the most frequent apparent cause of OMS. In adults, paraneoplastic etiology is more common, most due to lung or breast cancers. In contrast to paraneoplastic OMS in infants and young children, whose tumors are biologically inactive and often benign, the tumors in adults are commonly malignant, often disseminated. OMS is a rare disorder: 1 per million individuals worldwide. It usually affects infants and young children, although it is also known to affect adults. The peak age in children is about 18 months, with very few diagnosed before 1 year, and a long tail out to about 5 – 6 years. Occurrence of opsoclonus in infants under 6 months old is quite uncommon, and opsoclonus in that age group, when isolated, is usually from another cause. OMS occurs in only slightly more girls than boys. It occurs in about 3% of all children with neuroblastomas. The diagnosis is clinical; there is no diagnostic test yet, as the antigen remains unidentified. The presence of the 'dancing eyes', the shock-like muscle spasms, and the impairment of gait, especially if accompanied by irritability, are highly reliable indicators of this syndrome. To detect a tumor in children, either a CT scan with oral and IV contrast or MRI with gadolinium of the neck, chest, abdomen, and pelvis need to be done. PET scanning is often done in adults with OMS looking for other occult tumors. In addition, a spinal tap to detect neuroinflammation is necessary. Besides routine tests for infection, recommended CSF studies include so-called "MS panel", to include oligoclonal bands (with paired serum sample), looking for antibodies secreted by B cells in the CSF. Also, lymphocyte subset analysis (flow cytometry) using immunophenotyping reveals an increased frequency of CSF CD 19+ B cells, which is an invaluable biomarker of OMS disease activity. Autoantibodies in some children with OMS have been detected in research laboratories, but commercial autoantibody testing is not cost-effective and best reserved for atypical cases.