A case of intermixed ganglioneuroblastoma with OMS improved neurologically by tumor resection

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[Introduction] Opsoclonus-myoclonus syndrome (OMS) is a rare neurological syndrome that characterized by rapid, involuntary, irregular conjugate eye movement, myoclonic jerking of the limbs and trunk and cerebellar ataxia. More than half of pediatric patients with OMS suffer from neuroblastoma. [case] 5-year-old girl, who were suffering with an acute onset of ataxia, difficulty in walking and sitting over a period of 8 months. Furthermore, she got a speech disorder and independence of excretion disabled. A right adrenal tumor with calcification of 4-cm size was pointed out by the abdominal CT scan after 7 months from appearance of these symptoms. Her brain MRI shows no abnormality and MIBG scintigraphy shows no metastasis. She was perfomed the tumor resection and lymph nodes dissection laparoscopically. Histological examination showed schwann cells on fibrolastic stroma. In these cells, ganglion-differenciated cells were observed. In some of these specimens, blastic foci were observed and diagnosed intermixed ganglioneuroblastoma. After the surgery, these neurological symptoms were dramatically improved. [Discussion] The etiology of OMS is thought to be immune mediated by an immune response directed against the tumor, and cross-reacting with central neuronal tissues. But the exact pathogenesis has been unclear. It have been reported that about a half of neuroblastoma with OMS suffer from some neurological symptoms still after the surgery. In this case, her motor disturbances were gradually improved after the tumor resection. It is important to follow up for other neurological symptoms such as cognitive or behavioral problem. [Conclusion] We present an unusual case of ganglioneuroblastoma with OMS, which symptoms improved after tumor resection.