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Short communication

Marked improvement in opsoclonus and cerebellar ataxia after the surgical removal of a squamous cell carcinoma of the thymus: A case report

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ABSTRACT

A 69-year-old man with rapidly evolving vertigo and ataxia was admitted to our hospital. He was presented with a dysarthric speech and chaotic eye movements, identified as opsoclonus. Neurological examination revealed limb and truncal ataxias and an inability to stand unless fully assisted. A chest CT scan revealed a mass at the anterior mediastinum, which suggested paraneoplastic neurological syndrome (PNS). However, an extensive search for anti-neuronal antibodies linked to cerebellar ataxia failed to find any autoantibodies, including cell surface autoantibodies. Subsequently, a total surgical removal of the thymic tumor was performed, leading to marked improvements in his signs and symptoms. The pathological findings by conventional and immunohistochemical examinations confirmed a squamous cell carcinoma of the thymus. Three months after onset his signs and symptoms improved and he was able to walk without support.

In contrast to thymomas, PNS is extremely rare in patients with thymic carcinoma. Previous reports have shown that neurological symptoms, similar to opsoclonus or cerebellar ataxia, deteriorated in cases of thymic carcinoma that could not be controlled. The present report indicates that early diagnosis and total removal of the rare neoplasm may increase the possibility of neurological recovery.

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1. Introduction

Thymic carcinomas are rare epithelial neoplasms arising from the thymus and are distinct from thymomas, based on the World Health Organization (WHO) classification [1]. Thymomas are associated with a variety of paraneoplastic syndromes, including myasthenia gravis (MG) [2] or pure red cell aplasia [3], whereas immune-mediated neurological disorders are rarely observed in patients with thymic carcinoma [4]. Indeed, only a few cases with myositis or MG have been reported in patients with thymic carcinoma [4–6].

On the other hand, subacute cerebellar ataxia or opsoclonus have rarely been associated with thymic carcinoma. A case of cerebellar ataxia in a patient with thymic endocrine carcinoma was linked with GAD antibodies, implicating an immune response to GAD expressed by neoplastic cells [7]. In addition, a patient with concomitant mixed type thymic carcinoma showing opsoclonus with cerebellar ataxia was reported, although anti-neuronal antibodies associated with paraneoplastic neurological syndrome (PNS) were not shown [8]. In two other cases, total surgical removal of the neoplasm could not be performed because of pleural metastasis or local invasion, leading to a deterioration of neurological symptoms.

To our knowledge, this is the first case of a patient with squamous cell carcinoma of the thymus showing opsoclonus with cerebellar ataxia, whose neurological symptoms markedly improved after total surgical removal of the rare neoplasm.

2. Case report

A 69-year-old man with rapidly progressive vertigo and ataxia was admitted to our hospital. The patient had been well until approximately 4 weeks before admission when he developed occasional diplopia, followed during the next weeks by an increasing vertigo, slurred speech and rapidly progressive ataxia. Consequently, difficulty in ambulation developed. There was no history of preceding infection.

On admission, neurological examination showed normal consciousness; orientation for time and place was normal. His speech was dysarthric and had titubation of the head when seated. His eye movements were continuous and chaotic; there were involuntary, irregular, rapid conjugate eye movements in all directions. Finger-to-nose and heel–knee testing revealed bilateral dysmetria. He was unable to stand unless given full assistance because of limb and truncal ataxia. Myoclonic seizure was absent, and motor and sensory examinations, including superficial and deep sensations, were also normal. The deep

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tendon reflexes were normal throughout, except decreased bilateral Achilles tendon reflexes. The results of the remainder of the examination, including autonomic function, were normal.

A complete blood count, serum protein, electrolytes, and renal and liver function tests were normal. Serum levels of thyroxine, thyroid-stimulating hormone, vitamin B1 and B12 were normal. Tests for human immunodeficiency virus, syphilis, antinuclear antibodies, anti-DNA antibodies, anti-SS-A/SS-B antibodies were also negative. In addition, serum GAD antibodies evaluated by radioimmunoassay were negative. Cerebrospinal fluid analysis showed a slight increase in protein level (72 mg/dL) without pleocytosis. Although a T2-weighted brain MRI showed areas of an increased signal in the periventricular white matter, thought to represent small vessel disease, no abnormal findings were found in the cerebellum or brain stem (Fig. 1A & B). SPECT also failed to detect abnormality in the cerebellum or brain stem. Radiologic evaluation by CT scan revealed a 2-cm mass at the mediastinum (Fig. 1C), where 18F fluoro-2-deoxy-glucose-positron emission tomography showed a hypermetabolic focus (Fig. 1D). No other abnormalities were seen in the thorax, abdomen or pelvic CT. PNS associated with an anterior mediastinal tumor was suspected, although the auto-antibodies (anti-HuD, Yo, Ri, CRMP5, Amphiphysin, Ma1, M2) were not detected by Western blot analysis (ravo PNS Blot; ravo Diagnostika GmbH, Freiberg, Germany). Furthermore, antibodies for cell-surface neuronal antigens (NMDAR, AMPAR, $GABA_BR$, mGluR1, mGluR5, LGI1, and Caspr2) were not detected either. Moreover, immunohistochemical studies were performed using rat brain processed to determine antibodies to neuropil of brain as reported previously [9]; however, immunohistochemical analysis failed to identify neuropil antibodies of our patient.

Because of his rapidly progressive symptoms, corticosteroid treatment was initiated with a 1000 mg of methylprednisolone given intravenously for three days, followed by a 60 mg of oral prednisone. After seven days of prednisolone treatment, the nausea, opsoclonus and ataxia improved slightly. Finally, the patient underwent surgical removal of the tumor in the anterior mediastinum because of suspected PNS.

The histopathological findings of the anterior mediastinal tumor included solid and invasive growth into surrounding adipose tissues without capsular formation, polygonal tumor cells showing focal faint keratinization, and distinct nuclear atypia with prominent nucleoli (Fig. 2A & B). Immunohistochemically, tumor cells were positive against anti-CD5 (Fig. 2C), bcl-2 and CD 117 (Fig. 2D) antibodies. From these findings, this case was diagnosed as a squamous cell carcinoma of the thymus, based on WHO classification.

After total surgical removal of the thymic carcinoma, his signs and symptoms were markedly improved and he was able to walk without support three months after onset.

3. Discussion

Here we report a patient with opsoclonus and cerebellar ataxia whose symptoms significantly improved following surgical removal of a rare squamous cell carcinoma of the thymus. No evidence of metastasis to any organs including the brain and improvement in opsoclonus with cerebellar ataxia following surgical removal indicates a paraneoplastic etiology in our patient. According to the internationally accepted diagnostic criteria for PNS [10], our patient was classified as having definite PNS.

Thymic carcinomas are very rare neoplasms that occasionally have been reported in association with polymyositis, dermatomyositis and MG [3–5]. With the exception of myositis or MG, only 2 cases with opsoclonus or cerebellar ataxia have been associated with thymic carcinoma [7,8]. One was a case with cerebellar ataxia associated

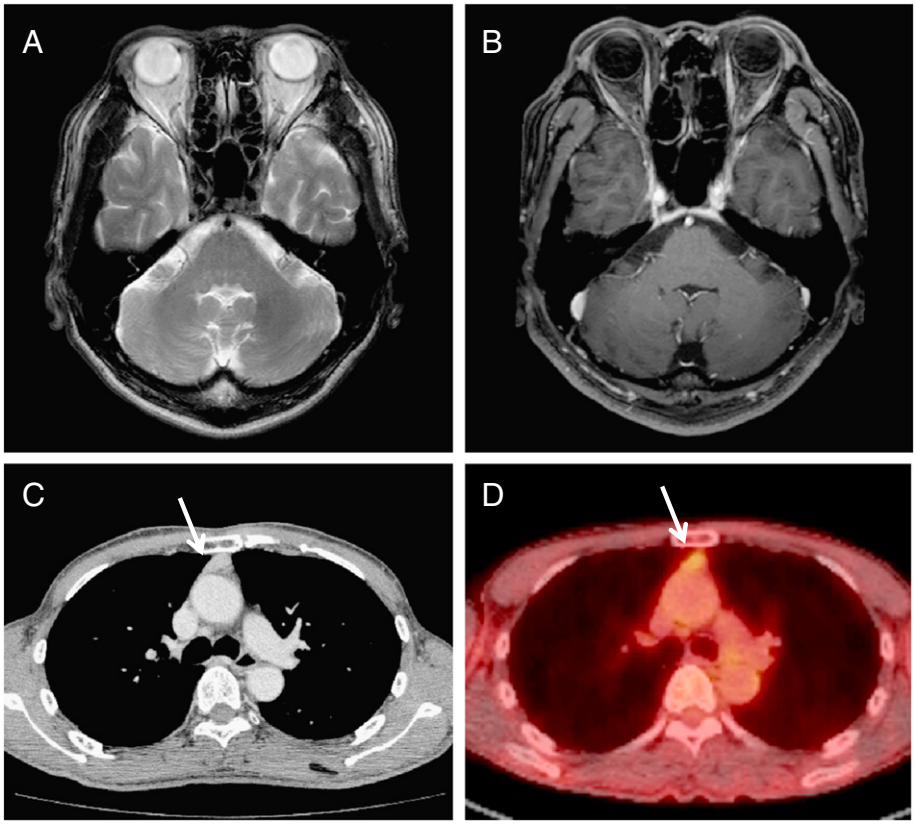

Fig. 1. Magnetic resonance imaging of the brain. Axial T2-weighted imaging (A) and T1-weight imaging after Gadolinium enhancement (B) show no abnormal lesion on the cerebellum or brainstem. Axial computed tomography of the chest shows an anterior mediastinal tumor (C, arrow). 18F fluoro-2-deoxy-glucose-positron emission tomography scan shows hypermetabolic focus on the anterior mediastinal tumor (D, arrow).

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