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Pharyngeal -Cervical -Brachial Variant of Guillain Barrè Syndrome Overlap with Subacute Combined Degeneration of Spinal Cord: Case Report at Department of Emergency

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Abstract: The pharyngeal-cervical-brachial (PCB) variant of Guillain–Barré syndrome is defined by rapidly progressive oropharyngeal and cervicobrachial weakness associated with areflexia in the upper limbs. Subacute combined degeneration (SCD) is an acquired myelopathy caused by vitamin B12 deficiency, and is a rare cause of demyelination of the dorsal columns of the spinal cord. We report the case of a patient evaluated at department of emergency (DEA) in our hospital, who received both diagnosis of PCB and SCD. This constitute a unique case in literature. Diagnosis of both diseases can be focused on history and neurological findings alone and it is important to promptly recognize them in order to start treatment.

Keywords: pharyngeal-cervical-brachial, mielopathy, Guillain Barrè.

INTRODUCTION

The pharyngeal-cervical-brachial (PCB) variant of Guillain–Barré syndrome is defined by rapidly progressive oropharyngeal and cervicobrachial weakness associated with areflexia in the upper limbs [Wakerley, B.R. *et al*., 2014]. The PCB variant of GBS is rare (0.07 - 0.25/100,000) [Emilia-Romagna Study Group, 1998]. Antecedent infection with CMV or Campylobacter Jejuni is often found with anti-GM2 antibodies being a hallmark of this variant [Yuki, N. *et al*., 1998]. Further diagnostic criteria for PCB variant include the presence of diaphragm weakness, relative sparing of lower limb strength and tendon reflexes, minimal or no sensory deficit, raised CSF protein, and generally axonal neuropathy on nerve conduction studies [Wakerley, B.R. *et al*., 2014; Nagashima, T. *et al*., 2007]. In most cases, patients experience a complete recovery [Wakerley, B.R. *et al*., 2014; Ropper, A.H. *et al*., 1991]. Subacute combined degeneration (SCD) is an acquired myelopathy caused by vitamin B12 deficiency, and is a rare cause of demyelination of the dorsal columns of the spinal cord [Timms, S.R. *et al*., 1993]. SCD affects the posterior columns and the corticospinal tracts and is characterized by swelling of the myelin sheaths and a patchy myelopathic spongy vacuolation of the affected regions of the cord [Renard, D. *et al*., 2009; Metz, J, 1992]. Neurologic symptoms of vitamin B12 deficiency are paresthesias, diminished proprioception and vibration sensation, motor weakness, clonus or hyperreflexia, areflexia, autonomic dysfunction, gait disturbance, intellectual or behavioral impairment, and impaired visual acuity [Healton, E.B. *et al*., 1991]. The most frequent neurologic manifestations are the SCD of the spinal cord and polyneuropathy [Healton, E.B. *et al*., 1991].

MRI findings in SCD can be diagnostically extremely helpful. MRI shows a very typical pattern with T2 hyperintense signal alterations usually confined to the posterior columns, which may involve the lateral columns and rarely brainstem [Srikanth, S.G. *et al*., 2002].

MATERIALS AND METHODS

We report the case of a 50 years-old Caucasian man who was evaluated at our department of emergency due to progressive worsening, in about 10 days, of upper limb weakness, dysarthria, dyspnea, mixed dysphagia. His medical history was positive for epilepsy from infancy treated with valproic acid 500 mg bid, and neurodevelopmental delay. He is a metal worker. Family history was unremarkable, especially for genetic and autoimmune disorders. In the last two months he denied respiratory or gastro intestinal infections, he didn't receive vaccines and he didn't travel (due to SARS-COV2 pandemia restrictions). He performed blood exams which resulted all normal, brain compute tomography (normal), lumbar puncture which revealed albumin-cytological dissociation (73 proteins upper normal value 45 mg/dl, 0 cells), with normal film array and cerebrospinal fluid cultures. Neurological examination showed bilateral facial palsy grade V House -Brackmann scale, dysarthria, upper arm weakness expecially proximal (grade 1/5 medical research council – MRC scale), cervical flexors weakness (3/5 MRC), mild lower arm weakness

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(4/5 MRC). MRC sum score resulted 26/40; Hughes scale score was 3. Upper limb reflex resulted all absent, while at lower limbs were bilaterally brisk. He didn't complain of positive or negative sensory symptoms. He had mild difficulty in gait (but was able to walk without aid), without ataxia. Spontaneous fasciculations were present at both deltoids and pectoral muscles. Eye movements and pupillary light reflex were normal. Body temperature and vital parameters resulted all normal; body mass index was 34.92 and he denied diarrhea, hematochezia or steatorrhea, such as alcohol intake. He was catheterized due no neurological bladder. He was promptly in-hospitalized and intravenous immunoglobulins were administered (0.4 g/kg for 5 days). Electromyography and electroneurography showed motor axonal-demyelinating neuropathy with prominent upper limbs involvement (prolonged F-waves latencies, marked reduction of compound muscle action potential amplitude registered from hand muscles). He also performed spine magnetic resonance imaging (MRI) showing he presence of posterior cords hyperintensity from C3 to D1 at T2 weighted sequences; highly suspicious for myelopathy (subacute combined degeneration of spinal cord) (images 1, 2). Vitamin B12 values resulted normal, while folic acid acid levels resulted lower than normal; consequently supplementation with folic acid was started. Screening neuropathy exams resulted all normal. Repeated nasopharyngeal swab resulted negative for SARS-COV2 infection. At day 1 from in-hospitalization muscle strength was 1 in the neck flexors, 1 in distal and 0 in proximal muscles of the upper limbs. Spontaneous fasciculations were present in all four extremities, expecially in upper limbs. parenteral nutrition had Parenteral nutrition had to be started due to persistent dysphagia. Laboratory tests including glucose, liver and muscle enzymes and electrolytes, seral protein levels were normal. Glycolipid antibodies couldn't be measured due to absence of laboratory testing. He was discharged to rehabilitation after 10 days. Neurological examination showed improvement in upper limb movements (muscle strength was 2 in the neck flexors, 3 in distal and 2 in proximal muscles of the upper limbs), bilateral facial dyplegia (House-Brackmann scale grade IV at discharge). He is still in-hospitalized in rehabilitative department of our hospital.

Diagnostic criteria for the pharyngeal-cervical-brachial (PCB) variant of Guillain-Barré syndrome (GBS) are:

Features required for diagnosis
Relatively symmetric oropharyngeal weakness AND neck weakness AND arm weakness AND arm areflexia/hyporeflexia*.
Absence of ataxia AND disturbed consciousness AND prominent leg weakness†.
Monophasic illness pattern AND interval between onset and nadir of oropharyngeal or arm weakness between 12 h and 28 days AND subsequent clinical plateau.
Absence of identified alternative diagnosis‡.

Features strongly supportive of the diagnosis
Antecedent infectious symptoms§.
Cerebrospinal fluid albuminocytological dissociation.
Neurophysiological evidence of neuropathy.
Presence of IgG anti-GT1a or anti-GQ1b antibodies.

*The clinical severity of each component may vary from partial to complete.

†The presence of additional features indicates overlap with other GBS variants as follows: ataxia AND ophthalmolpegia, 'PCB overlap with Fisher syndrome'; ataxia WITHOUT ophthalmoplegia, 'PCB overlap with acute ataxic neuropathy'; ataxia AND ophthalmolpegia AND disturbed consciousness, 'PCB overlap with BBE'. Leg weakness may vary considerable but oropharyngeal, neck and arm weakness should be more prominent. The absence of certain features indicates incomplete PCB as follows: upper limb weakness, 'acute oropharyngeal palsy'; pharyngeal palsy, 'acute cervicobrachial weakness (without pharyngeal palsy)'.

‡Including, but not limited to, brainstem ischaemia, myasthenia gravis and botulism.
§The presence of upper respiratory infectious symptoms or diarrhoea 3 days to 6 weeks before the onset of neurological symptoms.

Cerebrospinal fluid with total white cell count <50 cells/μL and protein above the normal laboratory range.

Our patient presented all strong features required for diagnosis and 2/4 features strongly supportive.

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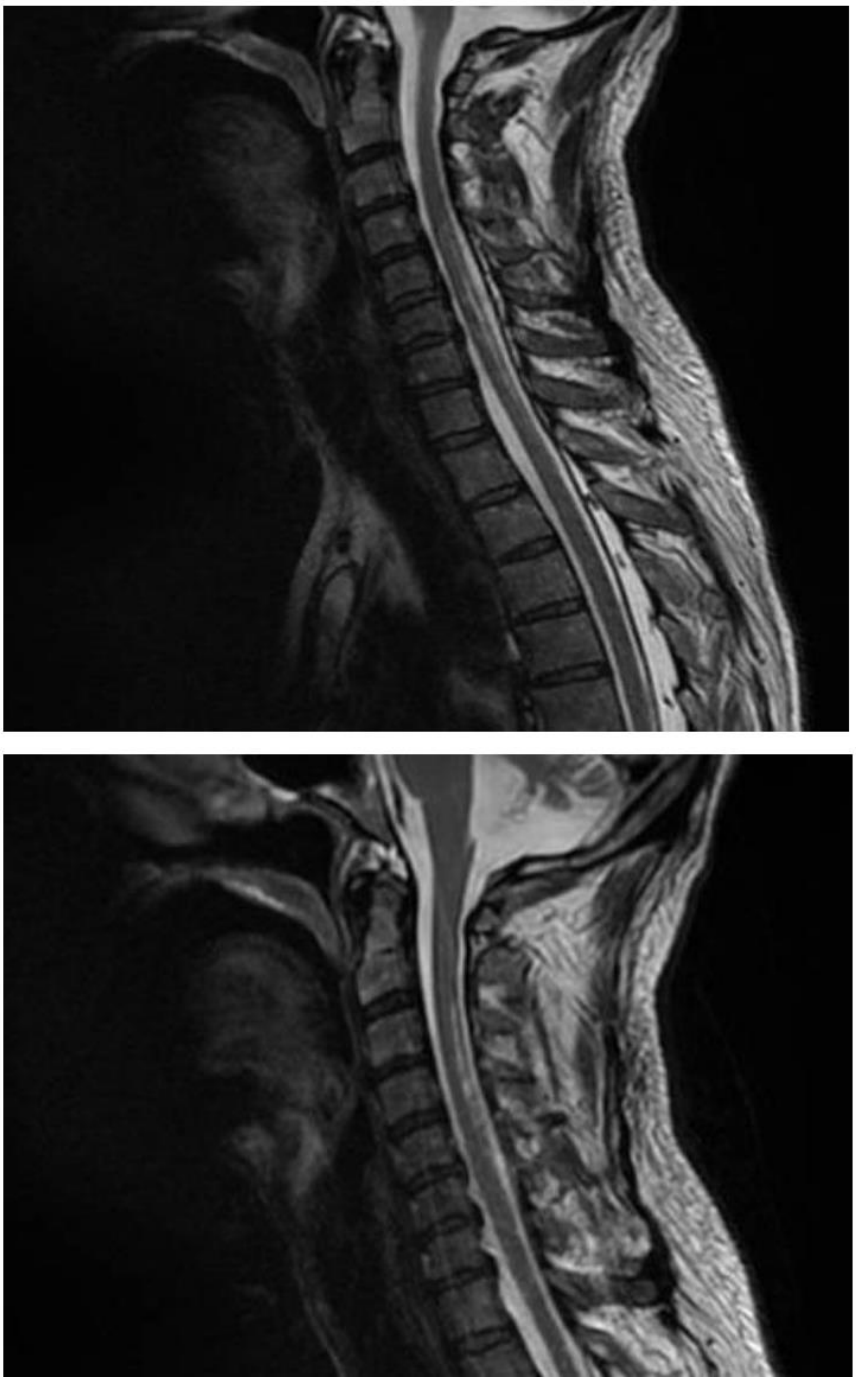

Figures 1,2: sagittal T2 MRI images showing hyperintensity of cervical posterior cord

DISCUSSION

Even if was difficult to perform neurological examination due to oligophrenia, the cardinal symptoms in our patient were the weakness of bilateral upper limbs, dysphagia, and dysarthria. Hypothesis of possible PCB was performed, promptly confirmed by neuroimaging, albumin-cytological dissociation and electromyography data (which allowed also to rule out other neurological diseases such as brain stroke, myasthenia gravis, botulism) but also prompt response to intravenous immunoglobulins administration.

He apparently denied other symptoms such as tingling, burning, and sensory loss of the distal extremities, ataxic gait that could have explained magnetic resonance imaging T2 signal abnormalities of cervical-dorsal cord, highly suggestive for SCD.

Serum folate deficiency was found while blood cell count, serum vitamin B 12 levels, lactate dehydrogenase and homocysteine resulted both

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normal. Human Immunodeficiency Virus infection was also excluded. Our patient will undergo testing for autoantibodies and gastroscopy to detect pernicious anemia. A possible causative role of valproic acid, assumed from infancy due to epilepsy, the pathogenesis of SCD, was ruled out after performing literature review [Geda, G. *et al*., 2002].

The possible coexistence of PCB and SCD has never been reported before in literature. Both diseases may be difficult to diagnose at the department of emergency: diagnosis can be made on history and neurological findings alone and it is important to promptly recognize and treat them.

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