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Clinical Case Seminar **A7(1-5)**

Dysphagia as initial manifestation of Guillan-Barrè Syndrome in a child

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Abstract

The occurrence of dysphagia in a child may be a sign of various pathological conditions and mainly gastrointestinal disorders; neurological causes are not very frequent, but they should be taken in to considerations. Etiological diagnosis is important for minimizing related complications. Here we report the case of a 6-year-old girl who was admitted to our Clinic with sudden onset weakness of the limbs and dysphagia. Physical examination revealed hypo-areflexia of both legs and arms and multiple cranial nerve dysfunction. Based on typical clinical course, laboratory investigations and electrophysiological studies, a diagnosis of Guillain-Barrè Syndrome (GBS) was assessed. A treatment with intravenous immunoglobulin (IVIG) was immediately started with a progressive recovery of motility and cranial nerve function.
An electrophysiological evaluation, performed one month after therapy start, showed slight improvement of neurological symptoms, in particular of the sensitive component. On the basis of our experience we suggest that a GBS should be suspected when dysphagia is associated with pain and ascending flaccid paralysis of the limbs, in order to prevent a severe complications such as respiratory failure.

Key-Words: dysphagia, Guillain-Barrè syndrome, flaccid paralysis

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Introduction

GBS is an acute, immune-mediated peripheral neuropathy, characterized by fast progressive muscle weakness and paraesthesia (1). Missed or delayed diagnosis and treatment can lead to progression of muscle deficits and inauspicious outcome for the involvement of the respiratory muscles (1).

Every year are affected by 1 to 4 persons per 100,000 adults and children, but more than two thirds of cases occur in childhood, especially among 4 and 9 years of age (2).

performed: cerebrospinal fluid examination revealed “albumin-cytological dissociation” (proteins 115 mg/dl, cells 1/mmc) pathognomonic of the syndrome.

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MRI with contrast of the brain and spinal cord was normal. Typical clinical course and CFS examination proved the diagnosis of GBS; therefore the child was treated with 0.4 g/kg/day of intravenous immunoglobulin for five days. She did not require respiratory support.

Electroneurography (performed two weeks after the onset of symptoms) revelaed a demyelinating neuropathy, mainly involving upper limbs and sensitive nerves.

Viral sierology per EBV and CMV were negative.

The patient presented a good response to treatment with a progressive recovery of motility and cranial nerve function; a second electroneurography examination, performed after 1 month from the previous one, showed a slight improvement, in particular of the sensitive component.

Case Report

A six year-old girl was admitted to our department for solid and liquid dysphagia and sudden onset weakness of the legs and the arms. 15 days before the onset of her symptoms, she had upper respiratory tract infection. The patient presented asthenia and lower limbs pain associated with difficulty to walk independently.

There was no history of exposition to toxic agents and her previous medical history was not significant. Mandatory vaccinations were complete. She reached our department transported by parents in stroller, unable to maintain the erect posture autonomously. On the admission she was apyrexial, fully conscious but very plaintive and irritable; vital signs were normal. She referred widespread hyperalgesia and burning sensation in the toes (especially in the right foot). Physical and neurological examination showed no sign of meningeal irritation but revealed **ataxia**, reduced motility and muscle power of the four limbs with areflexia in the lower limbs and hyporeflexia in the upper ones. Cranial nerve study showed a multiple dysfunction with mild ophthalmoparesis and limitation in bilateral eyes, inability to inflate the cheeks and to whistle, hypomobility of uvula and dysphagia, difficult tongue protrusion. She also presented dysarthria and hypophonia. There was no bladder and bowel incontinence. Examination of other systems did not show abnormalities. Laboratory tests revealed negative inflammatory markers with normal serum electrolytes and muscle enzymes (CPK, CK-mb, myoglobin). The character of worsening symptoms and the caudal-cranial extension in association with the abnormalities of neurological examination induced to suspect GBS. Therefore, after excluding expansive intracranial lesions with brain CT, a lumbar puncture was performed: cerebrospinal fluid examination revealed “albumin-cytological dissociation” (proteins 115 mg/dl, cells 1/mmc) pathognomonic of the syndrome. MRI with contrast of the brain and spinal cord was normal. Typical clinical course and CFS examination proved the diagnosis of GBS; therefore the child was treated with 0.4 g/kg/day

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Discussion

GBS is an acute, immune mediated inflammatory polyradiculo-neuropathy involving the peripheral nervous system (2) (3).

In about two thirds of patients it is preceded by a previous event, usually an upper respiratory tract, gastroenteritis or an infectious disease not well defined (4) (5). The most common pathogen involved is Campylobacter jejuni but several other bacterial and viral infections are associated (for example Haemophilus influenzae, Mycoplasma pneumoniae, Cytomegalovirus, Epstein–Barr virus and varicella zoster virus). There are also reported non-infective associations (6).

Causative agent triggers an immune response against gangliosides and glycolipids distributed along the myelin sheath (GM1-GD1b-GD1a) resulting in marked inflammation of peripheral nerves, demyelination and defective pulse propagation (7).

There are different subtypes of GBS; the principals are Miller Fisher syndrome (MFS), acute inflammatory demyelinating polyradiculoneuropathy (AIDP), acute motor axonal neuropathy (AMAN) and acute motor and sensory axonal neuropathy (AMSAN) (8).

The clinical presentation of GBS-related disorders is heterogeneous and the diagnosis may not be obvious at first.

Classic GBS begins abruptly with bilateral flaccid weakness and progressive motor deficit of the limbs. The weakness is often ascending and can involve the respiratory muscles (intercostal muscles and diaphragm) and cranial nerves with possible rapid progression to respiratory failure. Involvement of facial and oropharyngeal muscles is described in about 50% of cases and it can be the initial manifestation of the syndrome. In particular one third of patients require mechanical ventilation at some point during the disease. Impairment of respiratory muscles and dysphagia are the main factors that influence morbidity and mortality and therefore require close monitoring in the intensive care unit (9).

Oculomotor dysfunction, ataxia and areflexia are triad symptoms of the MFS.

The progression is fairly quick and fifty percent of patients overtake clinical nadir by 2 weeks (7).

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Symptoms (progression of motor deficit, relative sensory symptoms) are essential for the diagnosis supported by laboratory-instrumental tests; in particular the examination of the CFS showing typical albuminocytological dissociation (high protein, normal cell count).

Nerve conduction studies shows demyelinating finding like temporal dispersion, significantly slow conduction velocities and prolonged distal and F-wave latencies (8).

Although these studies should be performed early in suspected GBS, they are often non-diagnostic in the first week and should not be relied upon, or delay treatment, if there is a high index of suspicion for GBS. Repeat CSF and nerve conduction studies after the first week are therefore invaluable if there is any doubt over the diagnosis. Serum antiganglioside antibody testing can help only to support the diagnosis (7).

However the most important early investigation is neuroimaging to exclude an inflammatory, ischaemic or structural cause for weakness.

Patients with the confirmed diagnosis of GBS will be treated with plasmapheresis (in adult population) or intravenous immunoglobulin (IVIG).

IVIG is the treatment of choice of these patients to the typical dose of 0.4 g/kg/day for 5 days, although according to some evidences a total dose of 2 g/kg/day for 2 days, would be equally effective.

The majority of patients presents a gradual improvement in weeks or months, but 10 to 20% of these cases are complicated with a disabling motor deficit overall and about 3% of patients dies.

Our patient presented an important and progressive subtypes of GBS complicated by dysphagia and involvment of multiple cranial nerves, which responded quickly to treatment with IVIG.

A recent study shows that swallowing impairment is an important predictor of intubation in patients with GBS (9): fortunately in our patient it was not necessary because she promptly improved after IVIG treatment.

On the basis of our experience and the review of the current literature we emphasize the importance of a systematic clinical evaluation of swallowing, eventually combined with an evaluation of tongue protrusion strength, along with the usual assessment of neurological and respiratory function, to determine the severity of the GBS (9) and to start a useful therapy .

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