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Pharyngeal-cervical-brachial variant of Guillain-Barré syndrome: a rare cause of acute bulbar dysfunction in children

Variante faringo-cérvico-braquial da síndrome de Guillain-Barré: uma causa rara de disfunção bulbar aguda em crianças

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ABSTRACT

Aims: To report a case of pharyngeal-cervical-brachial variant of Guillain-Barré syndrome, which is characterized by rapidly progressive bulbar palsy with upper limb, neck and oropharyngeal involvement. It is a rare disorder in childhood and most cases have been described in adolescents.

Case Description: A seven year-old-boy presented with dysarthria, hoarseness, dysphagia, facial diplegia and bilateral progressive upper limb weakness. These symptoms started two weeks after a gastrointestinal infection. Nerve conduction studies were compatible with an acute demyelinating polyneuropathy in the upper extremities. Anti-ganglioside antibodies in the serum (anti-GT1a, GD1a, GQ1b) were positive and *Campylobacter jejuni* was isolated from stools. The patient was treated with intravenous immunoglobulin and needed ventilatory support during the first 12 days of admission. He was discharged at day 15 showing improvement of his neurological deficits. He fully recovered after eleven months of follow-up.

Conclusions: Although pharyngeal-cervical-brachial variant of Guillain-Barré syndrome is uncommon in children, it should be considered in a child with acute bulbar dysfunction because a timely diagnosis allows the early institution of therapeutic measures that can be lifesaving.

KEY WORDS: BULBAR PALSY, PROGRESSIVE; CHILDHOOD; GUILLAIN-BARRE SYNDROME.

RESUMO

Objetivos: Relatar um caso da variante faringo-cervico-braquial da síndrome de Guillain-Barré, que se caracteriza por paralisia bulbar rapidamente progressiva com envolvimento dos membros superiores, pescoço e região orofaríngea. É um diagnóstico raro na criança, ocorrendo a maioria dos casos em adolescentes.

Descrição do Caso: Um menino de sete anos de idade iniciou com queixas de disartria, disfonia, disfagia, diplegia facial e fraqueza muscular progressiva dos membros superiores. Estes sintomas surgiram duas semanas após uma infeção gastrointestinal. Os estudos eletrofisiológicos foram compatíveis com polineuropatia aguda desmielinizante nos membros superiores. Os anticorpos anti-gangliosídeo no plasma (anti-GT1a, GD1a, GQ1b) foram positivos e *Campylobacter jejuni* foi isolado nas fezes. O paciente foi tratado com imunoglobulina endovenosa e necessitou de suporte ventilatório durante os primeiros 12 dias. Teve alta no 15º dia com melhora dos sintomas neurológicos. Recuperou-se totalmente após 11 meses de seguimento.

Conclusões: Apesar da variante faringo-cervico-braquial ser pouco frequente em idade pediátrica, é um diagnóstico que deve ser considerado perante uma criança com disfunção bulbar aguda, pois a identificação precoce permite instituir rapidamente medidas terapêuticas que podem evitar a morte.

DESCRITORES: PARALISIA BULBAR PROGRESSIVA; CRIANÇA; SÍNDROME DE GUILLAIN- BARRÉ.

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INTRODUCTION

Guillain-Barré syndrome (GBS) is the most common cause of acute flaccid paralysis in healthy children. It affects both sexes, with male predominance, and occurs at any age, but it is rare in children under two years old. GBS is a heterogeneous syndrome that encompasses several variant forms. In its most common form it's characterized by an acute inflammatory demyelinating polyneuropathy. Patients usually present with sudden symmetric weakness in the lower extremities with diminished or absent deep tendon reflexes, that may ascend and involve the arms and respiratory muscles. These neurologic symptoms occur two to four weeks after a gastrointestinal or respiratory infection. *Campylobacter jejuni* is the microorganism most frequently identified as causing this previous infection.[1-3]

Etiology of GBS is unclear but the demyelinating form, especially when associated with an infection caused by *C. jejuni*, is believed to be immunologically mediated, and both humoral and cellular immune factors are involved. One-half of the patients may have autonomic dysfunction, in one-third there will be respiratory failure requiring ventilatory support, and facial weakness is described in 45 percent of all children with GBS.[1, 4]

Besides the demyelinating form, there are other variant forms with distinct clinical and pathological features of GBS: Miller Fisher syndrome is characterized by external ophthalmoplegia, ataxia and areflexia; Bicherstaff brainsteam encephalitis is a subtype of the Miller Fisher syndrome associated with consciousness disturbance; patients with Polyneuritis Cranialis have multiple bilateral cranial nerve involvement; and patients with the pharyngeal-cervical-brachial variant (PCB) have involvement of the upper limbs, neck and oropharynx. The PCB variant was first described by Ropper et al.[5] and has rarely been described in children.[6,7]

The authors present a case of a child who was diagnosed with PCB variant of GBS. This case illustrates that this is a rare cause of acute bulbar dysfunction and respiratory failure that needs a timely diagnosis.

This case report was approved by the ethics committee of the Hospital de Santa Maria (Refª DIRCLN-15JUL2013-0231) and the legal representative assigned the informed consent.

CASE DESCRIPTION

A seven-year-old boy was admitted to hospital with acute bulbar dysfunction and respiratory failure. He had no relevant medical history in his family. He was a preterm infant born at 28 weeks of gestation and needed intermittent ventilatory support after birth during the first fifty days of life. Two weeks before admission he had a self limited episode of bloody diarrhea. In this occasion he had eaten unpasteurized honey. Five days before admission he presented with fever, followed two days later by ineffective cough, dysarthria, hoarseness, dysphagia, facial diplegia and progressive bilateral upper limb weakness. He was observed in a first level hospital, and then transferred in the same day to a tertiary hospital after an aspiration pneumonia that caused respiratory failure.

On admission, he was alert and oriented. He showed signs of respiratory distress (central cyanosis, polypnea, tachycardia and hypoxemia) that motivated the need for ventilatory support. His eyes movements and pupillary light reflexes were normal. He had palsy of cranial nerves VII, IX, X, XI and XII which resulted in facial palsy, hoarseness, dysphagia and ineffective cough. Strength was decreased in the upper limbs. Muscle strength assessed using the Medical Research Council was graded as follow: neck flexors and proximal upper limb muscles, grade 4; distal upper limb muscles, grade 2; lower limb muscles, grade 5. Deep tendon reflexes were absent on the upper limbs and normal on the lower limbs. The plantar responses and sensibility were normal. He had no ataxia or tremor.

Complete blood count, serum electrolytes, liver function tests, and cerebrospinal fluid examination performed on the first day of hospitalization were normal. Cranial and spinal magnetic resonance imaging was normal on the second day. Nerve conduction studies also performed on the second day showed bilateral conduction block in the ulnar nerves, consistent with a demyelinating polyneuropathy; sensory nerve conduction function was normal. Anti-ganglioside antibodies in the serum (anti-GT1a, GD1a, GQ1b) were positive, and *C. jejuni* was isolated in the stools. Serologies were inconclusive because they were performed after administration of intravenous immunoglobulin.

He was diagnosed with PCB variant of GBS and was treated with intravenous immunoglobulin (IVIG) 400 mg/kg/day, for five days. He received antibiotics for two weeks to treat the aspiration pneumonia. By day 12, he no longer needed ventilatory support, and he was discharged from hospital at day 24. At that time he had mild facial palsy, palsy of XII cranial nerve, mild hoarseness, muscle strength grade 4 and hyporeflexia in the upper limbs.

After 11 months of follow up he had fully recovered and his neurological examination became normal. Anti-ganglioside antibodies in the serum remained positive at this time of follow-up.

DISCUSSION

PCB variant of GBS was first described in 1986 by Ropper.[5] The diagnosis of this variant was made based on specific clinical features, such as an acute progression of oropharyngeal, neck, shoulder, upper limbs and diaphragm weakness, bulbar palsy and preserved muscle strength in the lower limbs.[2,5] However, some patients were later described without diaphragm weakness and with slight weakness in the legs.[2-4] This shows that there is an overlapping of some variants of GBS, namely Miller Fisher syndrome, Bicherstaff brainsteam encephalitis and PCB, and these conditions form a continuous spectrum.[8] In the PCB variant the bulbar palsy appears in the earlier stages of the disease and it improves over a few months.

In the present case, symptoms related to the bulbar palsy and the involvement of the upper limbs and diaphragm were noted on the first days of the disease. Muscle strength in the lower limbs remained always normal and there were no alterations of consciousness. These clinical features were compatible with the PCB variant of GBS. Others conditions may present with acute and progressive bulbar palsy, such as myasthenia gravis, botulism, myositis, posterior fossa structural lesion, motor neuron disease, opercular syndrome and idiophatic cranial polyneuropathy. The neurologic examination, clinical course and workup exams allow the correct diagnosis.[1,8]

PCB variant is a rare disorder in childhood. The majority of the cases have been described in adolescents.[9] This patient is younger than those described previously. Generally this condition is preceded by an infectious illness of the respiratory or gastrointestinal tract. In PCB variant, *C. jejuni* and *cytomegalovirus* are the most common triggering agents.[2, 4, 9] Anti-ganglioside antibodies can be detected in patients with GBS and may be used as a marker for this condition, since specific nerve antigens are associated with the various variants of GBS.[4] In PCB variant, anti-GT1a IgG antibodies are the most common and are associated with a previous *C. jejuni* infection.[4,8] In MFS, BBE and PC anti-GQ1b IgG antibodies have been reported, which cross-react with anti-GT1a. Therefore, these specific antibodies may play an important role in the pathogenesis of the bulbar palsy in patients with PCB variant.[9] There are other anti-ganglioside antibodies that have been identified, but more rarely.[2] In this patient, there was a previous self limited gastrointestinal infection two weeks before the onset of symptoms.

Electrophysiologic studies can demonstrate a variety of abnormalities (including partial motor conduction block, slowed nerve conduction velocities, prolonged distal latencies) but can also be normal. In our patient nerve conduction studies indicated acute motor axonal injury in the arms without commitment of the sensitive fibbers. CSF examination of our patient was normal, but this is not uncommon in GBS, especially in the early stages.[7]

The diagnosis of PCB variant was supported by the clinical course, neurologic examination, neurophysiologic findings and the presence of anti-ganglioside antibodies in the serum (anti-GT1a, GD1a, GQ1b). Administration of IVIG and plasmapheresis are the main therapeutic options.[1] These two modalities have the same efficacy and safety profile; especially in more severely affected children but plasmapheresis may be difficult to perform in small children and involves placement of a central venous cathether.[4] The last systematic Cochrane review about the use of IVIG for GBS in children showed that IVIG hastens recovery compared to supportive care alone.[10] The authors state that it is low quality evidence and therefore more studies are needed, especially in mild disease and in patients whose treatment starts more than two weeks after onset of symptoms.

Referring to the long-term outcome of GBS, 85% of children will have an excellent recovery.[1] Mortality is 3% to 4% and is caused by respiratory failure or cardiac complications.[1] The prognosis correlates with the evolution of weakness and not with the severity of symptoms. Therefore, a better prognosis is associated with a slow and gradual evolution of weakness. Regarding to the PCB variant, the long-term outcome is better in children, since adult patients commonly have more prolonged disease.[4]

The impact on prognosis and long-term outcome resulting from administration of IVIG in children is still unclear. Vajsar et al.[11] established two factors that predict the long-term sequelae after administration of IVIG. The authors showed that children who are younger than nine years and who have progression to maximal weakness in less than 10 days are more at risk for a long-term deficit with greater residual weakness.[11] In the present case, the patient showed a favorable outcome after the administration of IVIG, with total recovery of his neurological deficits.

This case illustrates that respiratory failure and symptoms caused by bulbar palsy may dominate the clinical picture of the PCB variant of GBS. Although this is a rare condition among children, it can be rapidly fatal. When evaluating children for sudden onset bulbar palsy and limb muscle weakness we must include this disorder in the differential diagnosis. This approach allows for timely diagnosis and early institution of therapeutic and supportive measures.

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