

GULLAIN-BARRÉ SYNDROME AFTER VARICELLA-ZOSTER INFECTION

CASE REPORT

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In 1892, Sir William Osler noted an association between an acute febrile infectious illness and the later development of facial diplegia and limb weakness in some patients¹⁰. Indeed 50 to 70% of Guillain-Barré syndrome (GBS) in most series describe antecedent "viral" syndromes involving mainly the respiratory tract which precedes the symptoms of polyneuritis by one to three weeks, and a wide variety of specific viral entities preceding GBS has been described¹⁰. GBS following chickenpox has been seldom reported in the literature.

The authors describe a case of severe GBS after a prodromal varicella infection and make some considerations about this uncommon complication in a relatively benign exanthematic disease.

CASE REPORT

J.B., a 13 years-old young boy was admitted at 30th October 1986 complaining of progressive ascending paraparesis already involving the upper extremities and with dyspnea. Symptoms began for 4 days, and 11 days after a clinical picture of fever, malaise and vesicular skin eruptions compatible with chickenpox. At the time of admission he also complained of dysarthria and dysphagia, burning paresthesias of the lower members, sometimes painful, and diplopia. On examination the patient was anxious and dyspneic, he could not move his legs neither deglute his own saliva without great difficulty. There was a grade 3 paresis of the upper members, with global arreflexia and hypotonia. It was observed a superficial and profound anesthesia of the lower extremities, without a well-delimited upper level and the patient referred diffuse pain on his back irradiating bilaterally to the legs when he was passively moved. Painful dysesthesias improved well with the use of carbamazepine 600mg/day. It was noted a bilateral abducens paresis, bilateral peripheral facial palsy, with other cranial nerves normal. There were no fundoscopic alterations. On November 3rd his respiration worsened and he was intubated and connected to the respirator. A tracheostomy was performed at November 6th. During the first 3 weeks we observed the occurrence of episodic tachycardia, profuse sweat and salivation, and hypertension, which was controlled with the use of propranolol. No major arrhythmias did develop. An electrocardiogram was normal. A trial with dexamethasone was made for 4 days after what it was suspended as there was no response. Four days after being ventilated he developed a picture of herpes labialis which subsided in three days. He was maintained on mechanical ventilation until November 25th, when he resumed voluntary respiration without dyspnea, after a period of weaning off the respirator with intermittent mandatory ventilation (IMV). Hypertensive crisis subsided and antihypertensive drugs were withdrawn. At this time he was already tetraplegic, but could already deglute and talk with little difficulty. Bilateral facial and abducens paresis was still present. He was discharged on December 3rd, when he was bedridden and tetraparetic. He did not return for follow-up. The following laboratory tests in the serum were normal: aspartate and alanine aminotransferases, total bilirubin, alkaline phosphatase, creatine phosphokinase, lactate dehydrogenase, sodium, potassium, urea, creatinine, glucose. The hemogram at admission and during

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the hospitalization disclosed only leukocytosis with bastonetosis. Doehle corpuscles, without atypical lymphocytes nor anemia. A CT scan was normal. A CSF tap showed 27 red cells, 3 white cells (100% mononuclear), protein 92.0 mg/dl, glucose 64 mg/dl, chloride 118.6 mEq/l. Feces diluted to 15% were cultivated in cell cultures (VERO, Hep 2-C and RD) for isolation of enterovirus (polio, Coxsackie, Echo). No virus were isolated (Dr. Miguel Angez Rodriguez, Setor de Virus, Laboratório de Pesquisas Biológicas, Fundação de Saúde Caetano Munhoz da Rocha, Curitiba, performed the virus culture). The serological tests results are depicted in table 1.

COMMENTS

The Guillain-Barré syndrome is an acute or subacute, relatively symmetric ascending motor paralysis usually beginning in the lower members from which greater than 85 percent of patients obtain a full or functional recovery^{2,18}. The diagnosis is descriptive and rests upon recognition of the clinical picture plus other features including elevated cerebrospinal fluid (CSF) protein level (which can be normal at the beginning of the disease) plus light CSF pleocytosis or normal CSF white cell count, electrophysiological changes (marked slowing of conduction velocities, prolonged distal latencies, conduction block) and pathological changes (low-grade inflammation and demyelination-remyelination of the peripheral nerves)¹⁹. The presence of high-grade fever at the onset of the neuritic symptoms, transient bladder paralysis and severe sensory loss with pain may cause some confusion for the diagnosis of GBS¹⁵.

Anti-CMV (CF *)	1:4
Anti-CMV IgG (IF **)	0
Anti-CMV IgM (IF **)	0
Anti-EBV IgG	+
Anti-EBV IgM	0
Anti-poliovirus P1, P2, P3 (SN ***)	0
VDRL	1:2
Australia antigen (****)	0
Toxoplasmosis IgG (IF **)	1:6
Toxoplasmosis IgM (IF **)	0

Table 1 — Case. Serological tests: *, complement fixation; **, immunofluorescence; ***, soroneutralization (paired samples with 20 days interval); ****, radioimmunoassay; CMV, cytomegalovirus; EBV, Epstein-Barr virus; VDRL, Venereal Disease Reference Laboratory.

In 3 to 5 per cent of patients with GBS the polyradiculoneuropathy becomes chronic or recurrent. An increased prevalence of human leukocyte antigen (HLA) histocompatibility types Aw30, Aw31, B8 Dw3A and glyoxalase 1 is found in these patients²⁷, what is not observed in patients with acute monophasic GBS^{1,27}. A great variety of specific infectious agents have been implicated as causative of GBS, either by direct isolation or by rising antibody titers. Examples of such agents include para influenza A2, herpesviruses (herpes simplex, herpes zoster, cytomegalovirus, Epstein-Barr virus)^{3,10,21}, Echo virus, Coxsackie virus, influenza A and B, hepatitis B virus⁵, mumps²⁴, measles^{13,23}, falciparum malaria^{14,26}, infectious mononucleosis¹¹, *Mycoplasma pneumoniae*^{15,16}, tularemia²², and syphilis²⁰.

Deficits in pain and temperature sensation in a stocking-and-glove distribution predict greater residual impairment, since they imply severe axonal injury¹⁸. Autonomic dysfunction is not uncommon in GBS and cardiac arrhythmia is a frequent cause of death in these patients. These conditions were well observed in our patient. Neurologic complications at the time or following primary varicella infection in nonimmunocompromised patients are uncommon. The me-

chanism of nervous system disease caused by herpesviruses is still controversial²⁸ and include a direct invasion of the neural parenchyma by the virus. However the evidence of viral particles in the central or peripheral nervous system during disease is lacking. That means that an immunological process, non-dependent of the presence of the virus, may be operative. Both the cellular and humoral components of the immune system have been implicated in the mechanism of demyelination in GBS^{7,17}. Some viral antigenic determinants would "trigger" an abnormal host immunological response genetically determined (eg. HLA system). Apart the acute polyradiculoneuritis other immunologically mediated diseases have been described after varicella-zoster infection and include cerebellar ataxia, optic neuropathy, other cranial nerve palsies³ and encephalomyelitis^{20,28}. A similar pathogenesis theory has been proposed for the nervous system post-infectious complications caused by measles and rubella virus²⁰. In 302 cases of GBS studied by Samantray et al.²⁶, only one (0,3%) was related to chickenpox. Indeed there are few reports in the literature, and from 1873 to 1972 only 20 patients were cited in the world literature³². In recent reports the age of the patients varied from 9 to 32 years old^{6,8,9,30}. In all of them there was complete recuperation and two cases^{8,9} developed severe arterial hypertension as observed in our case, which later subsided completely.

One could postulate that the superimposed herpes simplex (herpes labialis) infection in our case could lead to a more severe and prolonged picture of polyradiculoneuritis due to antigenic and ultrastructural similarities between varicella-zoster virus and herpes simplex virus³¹.

SUMMARY

The authors report a case of Guillain-Barré syndrome (GBS) following a varicella infection in a 13 year-old boy. During his admission he developed respiratory insufficiency and dysautonomic events, as well as a severe sensitive peripheral neuropathy. Some aspects related to the etiology and pathogenesis of GBS are discussed.

RESUMO

Síndrome de Guillain-Barré após varicela: relato de caso.

Os autores relatam caso grave de síndrome de Guillain-Barré após quadro inicial de varicela em rapaz de 13 anos. Como complicações este paciente desenvolveu quadriplegia flácida com insuficiência respiratória, hipertensão arterial, taquicardia e importante neuropatia sensitiva periférica (disestesia dolorosa). Comenta-se a relação entre quadros virais benignos e o desenvolvimento de polirradiculoneurite aguda, assim como o possível mecanismo imunopatogênico envolvendo fatores humorais e celulares.

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