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Case Report

CIDP and Achalasia: Two manifestations of a Disease or Coincidental Association

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Abstract

Chronic inflammatory demyelinating polyneuroradiculopathy (CIDP) is an immune mediated disorder characterized by progressive developing or relapsing symmetrical motor or sensory symptoms in more than one limb over a period of two months. Achalasia, as a primary esophageal motility disorder, is also characterized by increasing the tone of lower esophageal sphincter, absence or incomplete sphincter relaxation in response to swallowing, loss of esophageal peristalsis and rising intra-esophageal pressure. Herein, a case of CIDP, dysmotility-like symptoms, and achalasia is presented.

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Keywords ● CIDP ● achalasia ● dysmotility

Introduction

Chronic inflammatory demyelinating polyneuroradiculopathy (CIDP) as well as achalasia are rare diseases of uncertain etiology in childhood. The concomitant presence of both disorders may suggest a common etiology, because a coincidental association would be remarkably rare. In 1994 Firouzi et al. presented a case of Guillain Barre Syndrome (GBS) with achalasia.[1] Some authors have classified CIDP as a chronic form of GBS.[2] This study presents a patient with CIPD that was associated with achalasia.

Case presentation

A 12-yr-old boy, form South of Iran, with dysphagia and inability to walk, was referred to Nemazee Hospital, affiliated to Shiraz University of Medical Science, Shiraz, Iran. The patient was well until two weeks before his first admission, when he developed multiple erythematous rash on his face and neck after a viral infection. His condition gradually deteriorated with weakness of lower and upper extremities followed by constipation, vague abdominal pain, and dysphagia. He had a negative family history and was not taking any drugs before.

The patient was alert and responsive on physical examination with body temperature of 37.2 °C, pulse rate of 110/min, and blood pressure of 105/63 mmHg. He had severe cachexia and soft distended abdomen with mild tenderness in all quadrants, and no signs of peritoneal irritation. Rectal examination did not show any fecal impaction. In neurologic examination he was orient and his gate was steppage with ataxia. His cranial nerves were intact and upper and lower extremity weakness was evident proximally and distally with motor power of 2/5. All deep tendon reflexes were absent with mute plantar reflexes.

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Abdominal, cremasteric, and bulbocavernous reflexes were also absent. Heel to shin and finger to nose tests were impaired, whereas, Romberg's sign was present. No sensory level was detected. He had no sense of pain, temperature, pinprick and position below his knee and elbow (socks and gloves pattern).

Initial laboratory investigations included hemoglobin of 12.1 g% with a white cell and platelet counts of 6,000/dl and 250,000/dl, respectively. The differential count showed 84% neutrophil, 7% lymphocyte, 6% monocyte, 1% band, 1% eosinophil and 1% basophile. Serum urea, creatinine and electrolyte levels and blood sugar, liver function tests and thyroid function tests (T3, T4, TSH, and T3-Rup) all were in normal range. Urinalysis was normal and negative for porphyria.

Cerbro spinal fluid analysis demonstrated a total cell count of 73/ml, containing 8 WBC, (88% lymphocyte, 12% segment), glucose concentration of 72 mg/dl with concomitant blood glucose level of 100 mg/dl and protein level of 131 mg/dl.

Plain abdominal x-ray and sonography were normal. Barium esophagography showed enlarged diameter of upper esophagus with decreased peristalsis in the lower part. “Bird’s beak” appearance was remarkable in favor of achalasia. Esoghagoscopy showed a large mass of bezoar in the distal part of esophagus with partial obstruction of the lumen. Furthermore, the MRI of cervicothoracic and thoracolumbar did not reveal any abnormality. Although EMG was normal, nerve conduction velocity showed motor and sensory polyneuropathy. Sural nerve biopsy showed demyelination and axonal degeneration with T cell and macrophage infiltration. Muscle atrophy was also seen in muscle biopsy, which was secondary to neuropathy.

During hospitalization he developed drooling and inability to swallow and persistent abdominal pain and aggravated dysphagia. He was transferred to Pediatric Surgery Ward for jejunal tube insertion. Explorative laparatomy revealed two small perforations in rectosigmoid junction. Sigmoid colostomy and peritoneal lavage was performed.

The patient was discharged after performing multiple esophageal balloon dilatations. In outpatient follow-up, he was well for two months, but he was referred due to cachexia and bed sore. The patient then was admitted for hyper alimentation. During hospitalization, he had severe colicky abdominal pain, mild distention with infrequent bowel movement. After stabilization, his abdomen was re-explored and closure colostomy was done. His condition improved with no abdominal pain. Afterward he tolerated oral feeding with satisfactory weight gain; however, still he had dysphagia and had difficulty in walking.

Discussion

CIDP is an immune mediated disorder characterized by progressive or relapsing symmetrical motor or sensory symptoms in more than one limb, developing over at least two months.[3] It represents the most common acquired developing treatable chronic neuropathy in childhood.[4] Sometimes the initial presentation of CIDP may mimic GBS as in our patient who presented with ascending pattern of weakness. CIDP was found more frequent in males than in females and history of antecedent illness or vaccination is present in about half of the cases,[5] as was observed in our patient who developed CIDP following viral infection. Proximal muscles are involved more than distal ones with the absence of muscle stretch reflexes,[3] as seen in this patient who had symmetrical proximal and distal weakness with the absence of deep tendon reflexes.

Our patient experienced prolonged abdominal pain without any evidence of peritoneal signs; however, in explorative laparatomy, bowel perforation and peritonitis were apparent. Therefore, these patients may have peritonitis without any peritoneal irritation. In this regard, when the patient's condition deteriorates one should consider bowel perforation in prolonged abdominal pain with soft abdomen.

For the diagnosis of CIDP different methods may be used. NCV test is suitable for evaluating nerves in CIDP patients,[7] which demonstrates reduction in conduction velocity, partial conduction block or abnormal temporal dispersion, and prolong distal latency in two or more nerves.[4] The NCV test performed in our patient suggested demyelinating polyneuropathy.

The most characteristic histologic features on nerve biopsy of patients with CIDP are loss of myelinated axons particularly large ones and inflammation.[4] The inflammation is investigated by measuring the number of onion bulbs, G ratio (axon diameter/total nerve fiber diameter), and the density of myelinated nerve fibers.[4] In this patient, biopsy of sural nerve demonstrated severe neuropathy in myelinated nerves and their axons without seeing any evidence of regeneration. T cells and macrophages were present in epineurium.

The present patient had achalasia and bowel dysmotility. The most common cause of achalasia is idiopathic, however neurogenic, myogenic and hormonal theories are also suggested.[6]

Fewer that 5% of children with achalasia manifested symptoms before 15 yrs of age.[7] The most common symptoms of achalasia are vomiting, progressive dysphagia, and weight loss. In order to diagnose achalasia, chest X-ray, barium swallow and esophageal manometry are helpful studies.

In fact, CIDP and achalasia are rare diseases of uncertain etiology in childhood, which both were present in our patient concomitantly. Therefore, one may consider CIDP and achalasia as two manifestation of a disease or coincidental association.

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