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# Sarcoidosis and Gullain-Barré Syndrome

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*Abstract*

*A 35-year-old female presented with three days' history of aching discomfort in her back, chest, and ankles. She had also noticed increasing weakness of her legs and a week before admission had shown flu-like symptoms. Chest X-ray showed bilateral hilar and right paratracheal lymphadenopathy. Bronchoscopic biopsies revealed non-caseating granuloma. She was diagnosed with sarcoidosis and was given prednisolone. The patient developed facial palsy and rapidly progressive ascending paralysis beginning from the lower extremities on the third and fourth days after initial presentation, respectively. Analysis of lumbar puncture showed acellular fluid with a high protein content. EMG was consistent with diffuse sensorimotor demyelinating polyneuropathy. Thus, the diagnosis was Guillain-Barré syndrome (GBS) presenting with sarcoidosis. Intravenous immune globulin was given and prednisolone stopped. One month after initial presentation the patient was completely recovered and discharged on prednisolone therapy. If neurologic symptoms such as aching discomfort and weakness are the main complaints in patients with suspected or biopsy proven sarcoidosis, GBS should be suspected.*

*Key words*: Sarcoidosis; neurosarcoidosis; Guillain-Barré syndrome.

## Introduction

Sarcoidosis is a poorly understood granulomatous disease that involves the lung and intrathoracic lymph nodes in > 90% of patients (1, 2). Neurologic complications occur in approximately 5% of patients with sarcoidosis (3). Approximately 50% of patients with neurosarcoidosis present with neurologic symptoms at the time sarcoidosis is first diagnosed (4, 5). Guillain-Barré syndrome (GBS) is an acute idiopathic polyneuropathy that sometimes follows infective illness, or surgical operations. Until now only six cases of GBS in patients with sarcoidosis have been reported in the literature (6-11). We present a case of GBS presenting with sarcoidosis.

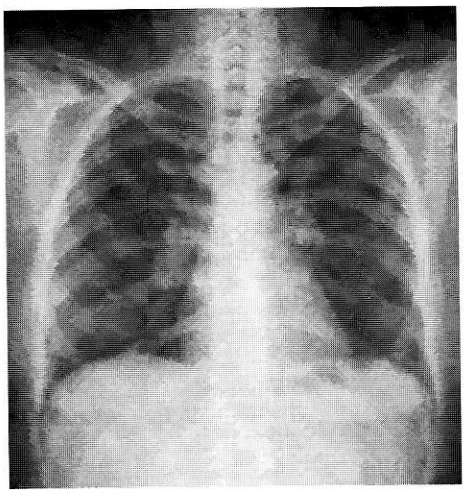


FIG. 1. — Chest x-ray: Bilateral hilar and right paratracheal lympadenopathy.

## Case report

A 35-year-old female was admitted with three days' history of aching discomfort in her back, chest, and ankles. She had also noticed increasing weakness of her legs, and one week before admission she had shown flu-like symptoms. Past history was normal. Physical examination was within normal limits. Investigations including complete blood count, whole blood chemistry, and spirometric pulmonary function tests were normal. Sedimentation was 12 mm/hour. Chest X-ray was abnormal with bilateral hilar lymphadenopathy and right paratracheal lymphadenopathy (Fig. 1). High resolution chest CT showed multiple mediastinal and bilateral hilar lymphadenopathies and acinar nodules on both upper

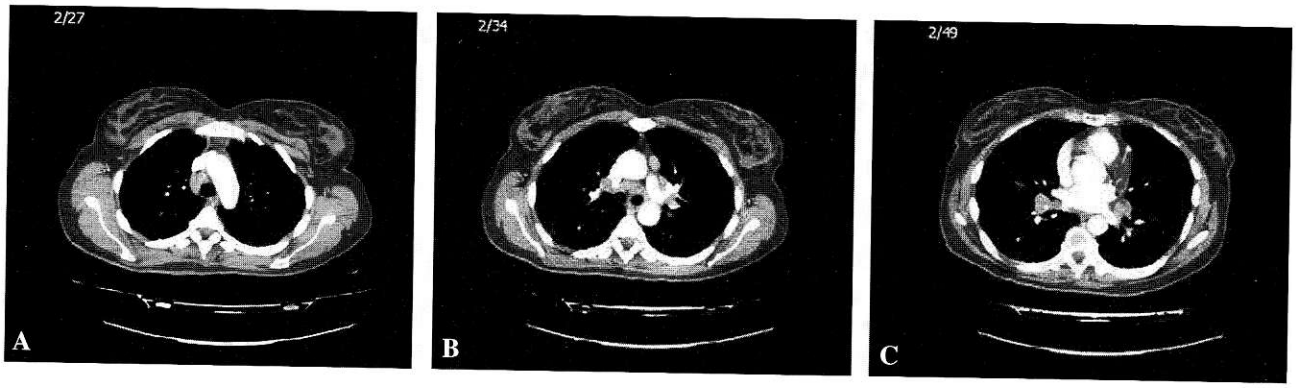



FIG. 2. — A. Chest CT : Right paratracheal lymphadenopathy; B. Chest CT: Mediastinal lymphadenopathies including precarinal and para-aortic lymhadenopathy; C. Chest CT: Bilateral hilar lymphadenopathy.

lobes (Figs. 2A, B, and C). Fiberoptic bronchoscopy revealed increased vascularization with cobblestone appearance on bronchial mucosa. Biopsies were taken from both mucosa and parenchyma, and histopathologic examination of the biopsies reported noncaseating granulomas including histiocytic giant cells and epitheloid histiocytes. Purified protein derivative (PPD) was negative. Autoantibody panel including p-ANCA,c-ANCA,anti-SS-B, anti-SS-A, anti-dsDNA, antiSm, antiScl70, RF, and AMA was negative. Thus, the final diagnosis was sarcoidosis and the patient was given prednisolone, po 40 mg/day. On the third day of treatment, however, she developed facial palsy. On the following day she developed rapidly progressive ascending weakness beginning from lower extremities. Cranial, cervical and thoracolumbar MRI, EMG, and lumbar puncture were performed after a neurology consultation. The MR sections were completely normal. Rapidly progressive ascending weakness beginning from lower extremities. Neurophysiology studies (electromyography and nerve conduction studies) showed absent F waves and absent H reflexes. Sensory nerve conduction studies reveal slowed conduction velocities. The sural sensory response was normal, while median and ulnar sensory responses were affected (sural sparing). Analysis of cerebrospinal fluid revealed acellular fluid with a high protein content (158 mg/dL). Final diagnosis was GBS and intravenous immune globulin (IVIG) was given and prednisolone stopped. After five days' treatment with IVIG the patient undertook physical therapy. Weakness improved dramatically and the patient began to walk. Prednisolone, po 40 mg/day, was begun one week later and the patient was completely recovered and discharged after 30 days' hospitalization.

## Discussion

Neurosarcoidosis is a diagnostic consideration in patients with known sarcoidosis who develop neurologic complaints and in patients presenting de novo with a constellation of findings consistent with the disease. Sarcoidosis can affect any portion of the central or peripheral nervous system. Common syndromes are cranial mononeuropathy, neuroendocrine dysfunction, restricted or generalized encephalopathy/vasculopathy (12) myelopathy/radiculopathy, communicating or noncommunicating hydrocephalus, acute aseptic meningitis or chronic meningitis, and peripheral neuropathy (13). Peripheral neuropathic presentation includes a mononeuropathy, mononeuritis multiplex, and generalized sensory, small fiber sensory, sensorimotor, and motor polyneuropathies (13). The symptoms can be acute, subacute, or chronic; electromyography usually reveals an axonal neuropathy. Differential diagnosis is important for differentiating neurosarcoidosis from other etiologies such as GBS, because corticosteroid treatment may be detrimental.

The acute immune-mediated polyneuropathies are classified under the eponym GBS. Most often, GBS presents as an acute monophasic paralysing illness provoked by a preceding infection. The cardinal features of GBS are progressive, fairly symmetric muscle weakness accompanied by absent or depressed deep tendon reflexes. Patients usually present a few days to a week after onset of symptoms (14). There is often prominent severe pain in the lower back, which is usually atypical for patients with sarcoidosis. (as in our case, who had an aching dyscomfort in her back). The typical finding with lumbar puncture is an elevated cerebrospinal fluid

protein with a normal white blood cell count, this finding is called albuminocytologic dissociation, and is present in 80 to 90 per cent of patients at one week after onset of symptoms. Clinical neurophysiologic studies (electromyography and nerve conduction studies) show usually evidence of an acute polyneuropathy with predominantly demyelinating features. The findings are prolonged or absent F waves and absent H reflexes. Sensory nerve conduction studies reveal absent responses or slowed conduction velocities. Typically, the sural sensory response is normal, while median and ulnar sensory responses are affected (sural sparing) (15-16). These features were seen in neurophysiologic studies in our case. Acute polyneuropathies that may mimic GBS include those due to acute arsenic poisoining, vasculitis, Lyme disease, tick paralysis, porphyria, sarcoidosis, leptomeningeal diasease, paraneoplastic disease, and critical illness. The combination of data from the clinical setting, laboratory testing, particularly electromyography with nerve conduction studies, and cerebrospinal fluid analysis, are usually sufficient to rule out these other causes of polyneuropathy.

Only six cases, of GBS occuring in patients with sarcoidosis have been reported in the literature (6-11). The patients were young (only one patient was 51 years old) and predominanly male (five male and one female). Our case was a young female. Time relation between GBS and sarcoidosis was so different that clinical picture of GBS began at two months prior to sarcoidosis, or at the same time with sarcoidosis, or one year after sarcoidosis. In our case, clinical picture of GBS was diagnosed at the same time with sarcoidosis. Usual diagnostic methods for GBS were; flu-like history, clinical findings (aching pain, acute facial palsy and rapidly progressive ascending weakness beginning from lower extremities), CSF results (acellular fluid with a high protein content), and neurophysiologic studies (electromyography and nerve conduction velocities). Since only six cases were reported and standard treatment was not present, each cases were treated differently such as prednisone, and mechanical ventilation. Our case was the first patient to whom IVIG was given. We stopped prednisone and gave IVIG and physical therapy and the patient improved rapidly and discharged after 30 days of hospitalization. Prognosis was also heterogenous changing from spontaneous recovery, to need for mechanical ventilation, and to death due to respiratory failure, (one patient died due to respiratory failure (9)). Our case had a good prognosis with complete recovery.

It seems likely that the clinical picture of our patient was due to GBS in view of the past history of flu-like illness, neurophysiologic study findings, albuminocytologic dissociation in cerebrospinal fluid, rapid onset of neurologic symptoms on third day of prednizolone treatment, and rapid resolution of the symptoms after IVIG, which a modifying therapy for GBS as plasma exchange and is as effective as plasma exchange for the treatment of GBS (17).

In conclusion, if neurologic symptoms such as aching dyscomfort and weakness (facial and/or ascending weakness beginning from lower extremities) are chief complaints in patients with suspected or biopsy proven sarcoidosis, and especially there is a history of flu-like illness within two-three weeks, GBS should be kept in mind and neurophysiology studies (electromyography and nerve conduction studies) should be done as early as possible for diagnosis because its treatment differs from that for sarcoidosis. And IVIG may be given to the patients with severe clinical picture. But further cases and studies are needed to clarify this.

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