

# Guillain–Barre´ syndrome with bilateral facial palsy with arachnoid cyst

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## ABSTRACT

This case report describes the findings of a 10-year-old female child admitted with fever and headache for 3 days along with several episodes of vomiting and generalized seizures for 1 day. She regained consciousness 50 minutes after admission with initial treatment, but developed muscle weakness, followed by tetraplegia with areflexia, bilateral facial palsy and respiratory insufficiency. The nerve conduction study (NCV) had evidence of demyelinating neuropathy and the cerebrospinal fluid (CSF) showed albumino-cytological dissociation. These neurologic findings were consistent with the diagnosis of Guillain-Barré syndrome. The patient was treated with immunoglobulin and other supportive cares. Mechanical ventilation was started on day 3 of hospital admission in face of respiratory insufficiency. Recovery, however, was relatively quick and extubation was possible on 6<sup>th</sup> day. Work up for CNS infection was non-contributory. MRI of brain done during the course, revealed an arachnoid cyst in right temporal region which might have contributed to the episode of unconsciousness, vomiting and convulsion. After two weeks of hospitalization, the patient was discharged from the hospital on wheel chair with satisfactory improvement of powers of facial and limb muscles. When the patient was seen at the outpatient department four weeks after hospital discharge, she was able to walk with help, with normal facial nerve function on both sides. GB syndrome presenting like meningitis; bilateral facial palsy; and co-presence of an arachnoid cyst makes this case worth sharing.

**Key Words:** Guillain-Barré syndrome, bilateral facial palsy, arachnoid cyst.

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## INTRODUCTION

Guillain-Barré syndrome (GBS) primarily involves motor, but may involve sensory and autonomic nerves also. It affects people of all age groups and is not hereditary. Most patients have a demyelinating variety of neuropathy, but primary axonal degeneration is documented in few cases.<sup>1</sup> The overall incidence rate of GBS is 1-2 per 100,000 in each year, but the rate is 0.8 per 100,000 each year in the people younger than 18 years. There is a male preponderance with male to female ratio of 1.5:1.<sup>2</sup> There may be an antecedent, presumably viral infection, triggering inflammation and demyelination. The patient may have bulbar involvement in about half of cases. Respiratory failure may occur. Some patients may have

facial nerve involvement. Some patients may also present with symptoms of viral meningitis or meningoencephalitis. CSF studies are important for diagnosis and may show an elevated protein content with the normal cell count.<sup>3,4</sup> Administration of intravenous immunoglobulin is essential in cases of rapidly progressive ascending paralysis.<sup>5</sup>

## CASE REPORT

A 10-year-old female child was admitted with fever and headache for 3 days along with several episodes of vomiting and generalized seizures for 1 day. The child was developmentally normal. She also had cough and cold for 2-3 days and there was similar history of cough and cold nearly 2 weeks prior to this present episode for

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which she received treatment from local physician. There was no history of chill and rigor, rash, dysuria, loose stool prior to this episode. She denied any history of trauma, intramuscular injection, regular drug intake heavy metal intoxication, recent vaccination, sera use, and injection in the gluteal region. The patient was drowsy on admission. On examination, she had grade II coma, pulse rate was 92/min, respiratory rate was 16/min, blood pressure was 124/78mm Hg and temperature recorded was 38°C. She had no neck stiffness and Kernig's sign was negative. The child was started on moist oxygen, intravenous fluid, antibiotics (ceftriaxone and amikacin), acyclovir and lorazepam. She regained consciousness 50 minutes after admission, but started complaining of weakness in both the lower limbs. In next 24 hours, weakness progressed rapidly to involve both the upper limbs also. Generalised hypotonia was noted. Lower limb power was 2/5 across all muscle groups; while power in upper extremity was 3/5 in all muscle groups. While she did give a history of 'pins and needles' sensation on both hands and feet, and stiff shoulders, she denied any numbness over the sites. She was aware of altered taste sensation. No cranial nerve palsy was noted at this point of time. Plantar response was bilaterally down but deep tendon jerks could not be elicited on any side in both upper and lower limbs. There was no bladder or bowel involvement at any point. Fundoscopy was normal. Examination of other major organ systems was non-contributory.

Initial laboratory work up revealed: a hemoglobin level of 11.8 gm%, total leukocyte count was 8300/mm<sup>3</sup> (neutrophil 79%, lymphocyte 15%, eosinophil 4%, monocyte 2%), platelet count was 2.98 lakh/mm<sup>3</sup>, peripheral smear was normocytic, normochromic and erythrocyte sedimentation rate was 15mm in first hour. The malarial antigenic tests were negative. Liver and renal function tests, serum electrolytes, blood sugar level, blood culture, Widal test, Mantoux test, chest x-ray, stool examination and urine studies were within normal limits. Cerebro-spinal fluid analysis showed a cell count of 5 per mm<sup>3</sup> (lymphocytes 100%), protein 66mg/dl, sugar 69mg/dl and ADA 9.8. CSF opening pressure was normal as measured with manometer attached to the lumbar puncture needle through a 3 way cannula. The CSF was also negative for syphilis (VDRL and hemagglutination), cysticercosis (ELISA and hemagglutination), pneumococci (latex), tuberculosis (acid-fast stain), meningococci (latex), H. influenza (latex), cryptococcus and HIV (ELISA).

Nerve conduction study (NCS) performed on 2<sup>nd</sup> day of admission demonstrated absent compound motor action potential (CMAP) in both peroneal and tibial nerves. Distal latencies were markedly prolonged in both median and ulnar nerves with reduced CMAP amplitudes and

conduction velocities. 'F' latencies were absent in all four limbs. Sensory nerve action potential (SNAP) were absent in both median, ulnar and sural nerves.

MRI of brain showed a cystic lesion in the right temporal region with extension to right side supra-sellar region with no mass effect on overlying cortical sulci (Figure 1). EEG was normal at this stage.

Fever was absent from 2<sup>nd</sup> day onwards. On day 3 of admission, patient suddenly developed respiratory difficulty, dysphagia, drooling of saliva and examination revealed bilateral lower motor neuron type facial palsy (House and Brackman Grade V) (Figure 2).

She had impaired gag reflex and right sided deviation of the uvula. She had partial loss of head control due to weakness of the sternocleidomastoid muscles bilaterally.

The patient was put to mechanical ventilator in the face of



**Figure 1:** Showing cystic lesion in the right temporal region with extension to right side supra-sellar region without mass effect.



**Figure 2:** Shows bilateral facial palsy.

impending respiratory failure. Intravenous immunoglobulin (IVIG) was given in the dose of 400mg/kg/day for 5 days. There was relatively rapid improvement of respiratory parameters in the next 2 days and the patient was extubated after 3 days of mechanical ventilation.

After consultation with neurology unit, a repeat lumbar puncture was done on day 10 of admission. Repeat CSF study showed a cell count of 7 per mm<sup>3</sup> (lymphocytes 100%), protein level was 120 mg/dl, sugar level was 72mg/dl (suggestive of albumino-cytological dissociation). Tests were negative for Lyme disease, Herpes simplex, *Borrelia burgdorferi* and Oligoclonal bands.

Her facial muscle function gradually improved, beginning with the right side. She was discharged on day 14, by then she was feeling significantly better and power in the upper and lower limbs were 4/5 and 3/5 respectively. Reflexes were normal in both the upper limbs but ankle and knee jerks in lower limbs were diminished. Advice was given to continue eye protection and return if there was any suggestion of worsening weakness or respiratory distress. She progressively improved and facial weakness had almost completely resolved on both sides by day 30 after discharge and she was able to walk with assistance when examined last at outpatient department. Patient was seizure-free in the meanwhile. She was ultimately referred to regional institute of neuro-surgery for consideration of any surgical intervention of the incidentally diagnosed arachnoid cyst.

## DISCUSSION

Children with GBS may present initially with muscle pain, involving the anterior and posterior thighs, buttocks, and lower back. This is followed by symmetric, progressive weakness of the extremities and facial muscles. The condition may be accompanied by autonomic dysfunction, headache, ataxia, and meningismus. The patient may have facial nerve involvement, which is often bilateral, may be found up to fifty percent of fatal cases.<sup>6</sup>

An arachnoid cyst is an intracranial benign, nongenetic developmental cyst which contains spinal fluid and occurs within the arachnoid membrane.<sup>7</sup> The exact mechanism of formation during embryogenesis is still unclear. Postulated mechanisms of the enlargement of these cysts include secretion by the cells forming the cyst walls, a one way valve, or liquid movements secondary to the pulsations of the veins. The most common location of arachnoid cysts in the pediatric population is in the Sylvian fissure/middle cranial fossa.<sup>8</sup> The cysts may be asymptomatic or may present with headache, seizures, and hydrocephalus. Signs and symptoms of raised intracranial pressure and specific

features of neural compression may be present. The child may have developmental delay in 10 per cent cases. Suprasellar cysts may have few additional manifestations like hydrocephalus due to obstruction to the normal CSF pathway, precocious puberty, head bobbing and visual impairment.

Complications of the cysts include progressive enlargement, intracystic or subdural hemorrhage and neuroendocrine dysfunction. Surgery is only indicated if there is obstructive hydrocephalus due to cyst or if neuroimaging shows mass effect, with compression of normal brain or brainstem structures. Posterior fossa cysts frequently require surgical intervention.<sup>8,9</sup>

GBS in the present case was diagnosed on the basis of clinical findings, NCS indicating demyelination and axonal degeneration, and albumino-cytological dissociation in CSF. Disease process evolved too rapidly, treatment decision was made without undue delay and recovery process also was very satisfactory leaving very little doubt about the diagnosis of the case. MRI spine with contrast to see anterior horn cell enhancement was not done in our case to reduce further cost of treatment as diagnosis was quite obvious. Etiology of initial presentation of headache, vomiting and repeated seizures was not clear. It was neither explained by CNS infection (work up was non-contributory for meningitis) nor by raised intracranial tension (ICT) (ICT was normal on both the occasion). Thus, we assumed that the subarachnoid cyst diagnosed incidentally might have contributed to those manifestations even though symptoms resolved without any specific measures taken for it. Suggestion was received from neuro-medicine department to observe the behavior of the cyst for some more time before embarking on any surgical intervention as patient was seizure free from few days prior to discharge. Patient, however, was ultimately referred to regional institute of neurosurgery for further opinion regarding treatment of the cyst.

## CONCLUSION

Co-presence of an arachnoid cyst in association with GBS and bilateral facial palsy is rare in literature. We, therefore, like to share our experience.

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