نحوه برخورد با دختر ۱۴ ساله با علایم فلج حاد پیشرونده اندامها در درمانگاه پزشکی خانواده

استاد راهنما: آقای دکتر شفیعی ثابت نورولوژیست وعضوهیات علمی گروه پزشکی خانواده

اراعه دهنده: دکتر محمدتقی فتاحپور کارورز پزشکی خانواده

Chief Complaint

- بیمار دختر ۱۴ ساله با ضعف پیشرونده اندام های تحتانی

Present Illness

- بیمار دختر ۱۴ ساله که ۲ هفته قبل از مراجعه دچار علایم URI شامل عطسه، سرفه و آبریزش بینی شده و بعد از آن دچار ضعف پیشرونده اندام های تحتانی و اختلال راه رفتن و پاراستزی و بی حسی نوک انگشتان دستها و پاها شده بود.
- ضعف پیشرونده در طی ۵ روز به عضلات تنه و بازو ها گسترش یافته و بیمار دچار خستگی عضلانی زودرس و ناتوانی در حفظ posture و از دست رفتن تعادل شده بود.
- همچنین علایم پاراستزی و بی حسی از نوک انگشتان به سمت دست ها و پاها گسترش یافته بود.

Present Illness

- علاوه بر اینها، بیمار دچار علایم افتادگی دوطرفه صورت، دوبینی، دیسفاژی و اختلال تکلم (دیس آرتری) شده.
- بیمار درد در ناحیه کمربند شانه، کمر و ران های هر ۲ پا ذکر میکرد که به صورت throbbing بوده.
- همچنین علایم احتباس ادراری و facial flushing (سرخی گونه ها) و palpitation نیز در طی هفته قبل مراجعه ذکر میکرد.

- PMH: Neg
- PSH: Neg
- DH: Neg
- AH: Neg
- FH: Neg

Physical Examination

- **General Appearance:**
- **دختر ۱۴ ساله بیدار، هوشیار و ارینته به زمان، مکان و اشخاص و به سوالات پاسخ میداد. ill, toxic نبود.**

- **Vital Signs:**
- **T: 37.2 / spo2:9۸% / RR: 24 / PR: 1۱0**

Physical Examination

- ملتحمه pale نبود.
- اسکلرا icteric نبود.
- افتادگی دوطرفه صورت مشهود بود.
- حرکات قفسه سینه قرینه بود. سمع ریه ها نرمال بود و صدای اضافه یا کاهش صدا سمع نشد.
- سمع قلب نرمال بود و s1و s2 بدون سوفل سمع شد.
- شکم نرم و فاقد دیستانسیون و تندرنس بود. هپاتومگالی یا اسپلنومگالی یا توده ای لمس نشد.

Physical Examination

- breathlessness در حین تکلم داشت.
- hyperemia در صورت مشهود بود. ضایعات پتشی، پورپورا و اکیموتیک مشاهده نشد.
- در معاینات عصبی:
- اختلال تکلم به صورت slurred speech ، تکلم آهسته و گاها نامفهوم داشت.
- رفلکس gag کاهش یافته بود.
- مردمک ها قرینه و midsize و reactive بودند.
- حرکات چشم های هر دو سمت آهسته تر از حالت عادی و دامنه حرکتی کاهش یافته بود.

Physical Examination

- Force اندام های تحتانی در distal هر دوطرف حدود ۴/۵ بود.
- Force اندام های تحتانی در proximal هر دوطرف حدود ۲/۵ بود.
- Force اندام های فوقانی distal هر دوطرف حدود ۴/۵ بود.
- Force اندام های فوقانی proximal هر دوطرف حدود 3/5 بود.
- رفلکس های اندام های تحتانی کاهش یافته بود.
- رفلکس plantar در هردو سمت کاملا مختل بود.
- معاینه حسی در اندام های تحتانی مختل بود.

Physical Examination

- معاینه finger to nose و heel to shin نرمال بود.
- Nystagmus نداشت.
- اختلال راه رفتن به صورت steppage gate و foot drop مشهود بود.
- قادر به انجام tandem gait نبود.

Definition

- Guillain-Barré syndrome
  - The acute immune-mediated polyneuropathies are classified under the eponym Guillain-Barré syndrome (GBS). GBS is one of the most common causes of acute, acquired weakness and is often provoked by a preceding infection. GBS may be complicated in some cases by respiratory failure and autonomic dysfunction.

Epidemiology

- GBS is the most common cause of acute flaccid paralysis in healthy infants and children
- GBS occurs worldwide with an annual incidence of 0.34 to 1.34 cases per 100,000 persons aged 18 years or less
- While all age groups are affected, the incidence is lower in children than in adults. The incidence increases by approximately 20 percent with every 10-year increase in age.
- Males are affected approximately 1.5 times more often than females in all age groups.

Signs and Symptoms

- The typical patient with GBS, which in most cases will manifest as acute inflammatory demyelinating polyradiculoneuropathy (AIDP), presents 2-4 weeks following a relatively benign respiratory or gastrointestinal illness with complaints of finger dysesthesias and proximal muscle weakness of the lower extremities. The weakness may progress over hours to days to involve the arms, truncal muscles, cranial nerves, and muscles of respiration.

Weakness

- The classic clinical picture of weakness is ascending and symmetrical in nature. The lower limbs are usually involved before the upper limbs. Proximal muscles may be involved earlier than the more distal ones. Trunk, bulbar, and respiratory muscles can be affected as well.
- Patients may be unable to stand or walk despite reasonable strength, especially when ophthalmoparesis or impaired proprioception is present. Respiratory muscle weakness with shortness of breath may be present.
- Weakness develops acutely and progresses over days to weeks. Severity may range from mild weakness to complete tetraplegia with ventilatory failure.

Cranial nerve involvement

- Cranial nerve involvement is observed in 45-75% of patients with GBS.
- Common complaints include the following:
  - Facial droop
  - Diplopia
  - Dysarthria
  - Dysphagia
  - Ophthalmoplegia
  - Pupillary disturbances

- Sensory changes
  - Most patients complain of paresthesias, numbness, or similar sensory changes. Sensory symptoms often precede the weakness. Paresthesias generally begin in the toes and fingertips, progressing upward but generally not extending beyond the wrists or ankles. Loss of vibration, proprioception, touch, and pain distally may be present.

  - Sensory symptoms are usually mild. In most cases, objective findings of sensory loss tend to be minimal and variable.

- Autonomic changes in GBS can include the following:
  - Tachycardia
  - Bradycardia
  - Facial flushing
  - Paroxysmal hypertension
  - Orthostatic hypotension
  - Urinary retention

Respiratory Complaints

- Upon presentation, 40% of patients have respiratory or oropharyngeal weakness.
- Typical respiratory complaints in GBS include the following:
  - Dyspnea on exertion
  - Shortness of breath
  - Difficulty swallowing
  - Slurred speech

DDX

- Chronic inflammatory demyelinating polyneuropathy
  - AIDP is a monophasic subacute illness that reaches its nadir within three to four weeks. CIDP continues to progress or has relapses for longer than eight weeks
- Other polyneuropathies
  - Acute polyneuropathies that may mimic GBS include those due to acute severe vitamin B1 deficiency, nitrous oxide-induced neuropathy, acute arsenic poisoning, vasculitis, Lyme disease, tick paralysis, porphyria, sarcoidosis, leptomeningeal disease, paraneoplastic disease, and critical illness.

DDX

- Spinal cord disorders
  - Acute myelopathies due to spinal cord compression or acute transverse myelitis can be confused with GBS, since reflexes can be depressed in the acute stage of spinal cord disease. Early bowel and bladder dysfunction and a sensory level point to a myelopathy. Imaging with spine MRI is usually helpful in diagnosing acute myelopathy by demonstrating a focal spinal cord lesion.
- Neuromuscular junction disorders
  - Diseases of the neuromuscular junction including botulism and myasthenia gravis can present with acute weakness, but without sensory signs or symptoms.

DDX

- Muscle disorders
  - Acute polymyositis, critical illness myopathy, and critical illness neuropathy can mimic GBS.

Work up and Diagnosis

1.Clinical Diagnosis:

- •GBS is generally diagnosed based on clinical grounds.

- •Basic lab studies (CBCs, metabolic panels) are normal but ordered to exclude other diagnoses and assess functional status.
- •Specific tests guided by patient history and presentation.
- •Check CBC, Electrolytes, LFT, CPK, ESR

2.Electromyography (EMG) and Nerve Conduction Studies (NCS):

- •Electrodiagnostic studies are the most specific and sensitive tests for diagnosis of GBS and establish the underlying pathophysiology as either demyelinating or axonal.
- • Abnormalities consistent with demyelination (delayed latencies, slowed conduction, etc.) support classic GBS.
- •Needle EMG may be normal initially; fibrillation develops later.
- •Normal studies don’t rule out GBS.

3.Pulmonary Function Monitoring:

- •Frequent evaluations needed to monitor respiratory status and ventilatory assistance.
- •Forced vital capacity (FVC) is very helpful in guiding disposition and therapy.
- •Negative inspiratory force (NIF) is a relatively easy bedside test. Normal is usually greater than 60 cm water. If the NIF is dropping or nears 20 cm water, respiratory support needs to be available.

4.Lumbar Puncture (CSF Studies):

- •Recommended during acute phase.
- •Characteristic CSF findings: elevated protein (>0.45 g/L) without increased WBC (albuminocytologic dissociation).

5.Imaging Studies:

- •MRI and CT scanning mostly help exclude other diagnoses (mechanical myelopathy).
- •MRI is sensitive, but nonspecific, for diagnosis. However, it can reveal nerve root enhancement and may be an effective diagnostic adjunct.

- Patients with GBS should be admitted to an inpatient setting for serial hemodynamic and neurologic monitoring and to guide therapy.

- We admit (or transfer) patients to an intensive care unit (ICU) setting who present with weakness that is severe or rapidly progressive and are at high risk for respiratory failure, those with signs of imminent respiratory failure, and those with autonomic dysfunction.

Treatment

- Competent intensive care includes the following features:
  - Respiratory therapy
  - Cardiac monitoring
  - Safe nutritional supplementation
  - Monitoring for infectious complications (eg, pneumonia, urinary tract infections, septicemia)

- Subcutaneous heparin was prescribed to prevent lower-extremity deep venous thrombosis (DVT) and consequent pulmonary embolism (PE).

- Immunomodulation
  - Intravenous immunoglobulin (IVIG)
  - Plasma exchange
- Physical, and speech therapy
  - Estimates suggest that approximately 40% of patients who are hospitalized with GBS require inpatient rehabilitation.

سطوح پیشگیری

Primordial Prevention

Primary Prevention

Secondary Prevention

Tertiary Prevention

Quaternary Prevention

Primordial Prevention

- **Health Education:** Promoting awareness about GBS risk factors (such as recent infections) and among the general population.

- **Lifestyle Modification:** Encouraging healthy habits (e.g., regular exercise, balanced diet) to reduce overall risk.

Primary Prevention

- **Vaccination:** Administering vaccines (e.g., influenza, COVID-19) to reduce infection-related GBS risk.

Secondary Prevention

- **Early Diagnosis and Treatment:** Promptly identifying GBS symptoms (muscle weakness, tingling) and initiating medical care.

- **Assessing Respiratory Function:** Assessments to detect breathing difficulties and provide necessary support as soon as possible.

Tertiary Prevention

- **ICU Management:** Close monitoring in an intensive care unit (ICU) for complications (e.g., breathing difficulties, blood pressure fluctuations).

- **Monitoring for autonomic disturbances**: Including changes in blood pressure and pulse rate, as well as respiratory, bowel, and bladder dysfunction.

Quaternary Prevention

Avoiding Unnecessary Procedures