

SELF ASSESSMENT ANSWERS

Shortness of breath

Q1: What is the diagnosis?

Varicella zoster virus (VZV) or chickenpox pneumonitis.

Q2: How might the diagnosis be confirmed?

Diagnosis can be made in most circumstances clinically. Confirmation and identification of the virus can be made in a number of ways using laboratory tests:

- **Polymerase chain reaction**—With high sensitivity (97%–100%) and specificity for viral DNA with rapid diagnosis time, this is a useful test, as it can be applied to most clinical specimens and is likely to replace older tests.¹
- **Serology**—Anti-VZV IgM can be used to detect active disease, while the presence of anti-VZV IgG is a marker of previous infection/inoculation.
- **Electron microscopy**—This is very sensitive but expensive and unable to differentiate between VZV and other herpes viruses—for example, herpes simplex virus (HSV).
- **Viral culture**—This is the gold standard for establishing the diagnosis, though VZV is more difficult to culture than HSV.
- **Tzanck smear**—An inexpensive “bedside” test from vesicular fluid with a sensitivity reported at 80%–100%.² Under light microscopy, the pathognomonic triad includes multinucleated giant cell formation, ground glass appearance of the nuclear chromatin, and nuclear contours that appear moulded together.
- **Direct immunofluorescence**—This utilises fluorescein-tagged antibody; more accurate than the Tzanck smear and able to differentiate between VZV and HSV.

Q3: What is the differential diagnosis of findings on the chest radiograph (see p 679)?

Miliary mottling is seen in a number of different disease processes; it describes a diffuse pattern of lung infiltration seen as numerous small opacities. Causes include: VZV pneumonitis, acute respiratory distress syndrome, miliary tuberculosis, fungal infection (invasive aspergillosis), extrinsic allergic alveolitis, fibrosing alveolitis, pneumoconiosis, and sarcoidosis.

Progress

The patient subsequently required admission to the intensive care unit where continuous positive airways pressure ventilation was used to maintain oxygenation. Over a course of 12 days he responded well to intravenous acyclovir and flucloxacillin. One month after discharge the patient was symptom-free, and a repeat chest radiograph demonstrated near complete resolution of infiltrates.

Discussion

VZV is a double stranded DNA virus of the alpha herpes virus family. Primary infection results in chickenpox and reactivation of the virus in later life causes shingles, or in severely immunocompromised patients, disseminated zoster.

VZV pneumonitis complicates between 2% and 10% of cases of adult chickenpox.³ The severity of VZV pneumonitis is highest in immunosuppressed persons, pregnant women, and in individuals who smoke.⁴

Microscopy of lung parenchyma in VZV pneumonitis shows focal necrosis, consolidation, a mononuclear infiltrate, and intranuclear inclusion bodies. VZV can be identified from vesicular fluid, respiratory secretions, or blood (as above).

Radiographic findings are diffuse, fluffy, reticular, or nodular infiltrates that can be rapidly progressive; this can give rise to the miliary pattern seen in the case presented. Pleural effusions can also occur. Radiographic abnormalities are often non-specific and more prominent during the peak of the rash and resolve rapidly with clinical improvement. Long term respiratory sequelae are infrequent in survivors, although small, diffusely scattered calcifications may persist on chest radiographs.

Treatment of VZV pneumonitis includes respiratory isolation until skin lesions heal, intensive respiratory support, administration of antiviral agents, particularly acyclovir and related compounds, antibiotics, and active and passive immunisation. Intravenous acyclovir is the drug of choice and needs to be started within 72 hours of symptom onset. Empiric antibacterial therapy is indicated in immunocompromised patients with VZV pneumonitis; respiratory failure may develop secondary to superimposed bacterial infection, with *Streptococcus pneumoniae*, *Staphylococcus aureus*, and *Haemophilus influenzae* commonly implicated.⁵

In conclusion, immunocompetent adults who contract VZV are at risk of developing life threatening complications including pneumonitis, and thus clinicians must have a high index of suspicion when attending these patients. Patients with respiratory failure will require admission for supportive measures and treatment.

Final diagnosis

Varicella zoster virus pneumonitis.

References

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Symptomatic bradycardia and postural hypotension

Q1: What is the initial work-up of syncope?

The first step in the work-up of syncope is to establish whether the patient truly had a syn-

copal episode. Syncope should be differentiated from vertigo, panic attack, hysterical attack, drop attack (in which consciousness is not lost), and disorders in which consciousness is lost such as metabolic disorders (hypoglycaemia, hypoxaemia) or epilepsy (box 1).

A thorough history, physical examination, and electrocardiogram are the cornerstones for diagnosis of syncope. History taking should focus on postural symptoms (postural hypotension), exertional symptoms (obstructive valvular disease, coronary ischaemia), situational symptoms (cough, micturition, defecation, food), predisposing circumstances such as during venepuncture (vasovagal) or turning the head while wearing a tight collar (carotid hypersensitivity syndrome), psychiatric history (panic attack, hysterical), history of structural heart disease (ventricular tachycardia), medications (antiarrhythmics, antidepressants), and family history (long QT syndrome, Brugada syndrome, hypertrophic cardiomyopathy). Cardiovascular examination (obstructive valvular lesions) and focal neurological deficit (cerebrovascular disease) may give pointers to the diagnosis.

An electrocardiogram may establish the diagnosis in 5% of patients with syncope. Routine use of basic laboratory tests is not recommended. Electrolytes, fasting glucose, serum levels of some medications, and cosyntropin test may be done if specifically suggested by history and physical examination. Computed tomography and magnetic resonance imaging should be avoided unless focal neurological features are present. Echocardiogram, stress test, and cardiac catheterisation may be required for exertional symptoms. In young patients with a structurally normal heart, Holter monitor and tilt table test may be required.¹

Q2: How can Arnold-Chiari malformation and syringomyelia be related to her symptoms?

Autonomic dysfunction may cause wide variations in blood pressure with supine hypertension and postural orthostasis. Associated symptoms of bladder dysfunction, abnormal sweating, or constipation may be found. Exaggerated responses to medications may be the first sign of an underlying autonomic disorder. Autonomic dysfunction has a varied aetiology. This includes peripheral neuropathies (most common cause), multiple system atrophy (associated involvement of pyramidal tract, cerebellar, and striatonigral pathways), and spinal cord disease. Autonomic disturbances due to involvement of sympathetic structures in the intermediolateral column of the spinal cord constitute a

Box 1: Conditions that mimic syncope

- Seizure.
- Cerebrovascular stroke/transient ischaemic attack.
- Hypoglycaemic episode.
- Hypoxaemia (asphyxiation).
- Vertigo.
- Hyperventilation syndrome/panic attack.
- Hysteria.

well recognised group of manifestations in syringomyelia.² These include Horner's syndrome, sweating abnormalities, and trophic and vasomotor changes in the limbs. Defective autonomic control of the heart has received less attention despite its potentially hazardous complications. Autonomic function as assessed by non-invasive cardiovascular reflexes and sympathetic skin response has been found to be abnormal in patients with syringomyelia. Orthostatic intolerance and postural tachycardia has been described in patients with syringomyelia.³ Fludrocortisone and β -blockers have been shown to lead to resolution of symptoms.

Q3: What are the final diagnosis, clinical features, and treatment of this disorder?

Intentional overdose of clonidine can explain symptomatic bradycardia with hypotension and its withdrawal probably lead to rebound hypertension. A psychological basis of cardiac symptoms occurs more often than is recognised. The final diagnosis of this patient is Munchausen's syndrome.

Typically patients with Munchausen's syndrome present with dramatic symptoms with inconsistencies in their complaints, demand extra attention when hospitalised, ignore hospital rules, and leave hospital against medical advice once the truth is discovered. They have a fair knowledge of medical terminology and may work in a related medical field, resist psychiatric evaluation, change doctors frequently, and travel great distances to seek medical attention. The diagnosis is often made by having unusual complaints or laboratory findings not explicable by any known disease or by discovery of incriminating items or self induced afflictions.

These patients evoke feelings of anger, contempt, hostility, and futility in physicians and medical staff. A supportive non-condemning environment must be maintained by the treating physician. Consultation with a psychiatrist must be obtained early on. Hospital administration, the risk management department, and ethics committee may need to be consulted to avoid lawsuits. A computer based central registry may help to identify these patients and prevent unnecessary hospital admissions and invasive procedures. Consistent support of caregivers and the therapist may reduce the need for factitious behaviour. Long term prognosis and follow up is not known in this subgroup of patients as they are prone to change their physician frequently once their true diagnosis is known.

Discussion

The diagnosis of syncope is often challenging. A thorough history and physical examination can help in the diagnosis of syncope in 45% of patients in which the cause can be determined. Although the yield of electrocardiography is low, it is inexpensive, risk-free, and may guide further evaluation and therefore is recommended in almost all patients with syncope. Unnecessary tests should be avoided in the workup of syncope. Syncopal episodes may be associated with bradycardia (box 2), tachycardia, or a normal heart rate (box 3).

The first turning point of the case was with the return of the patient with symptomatic bradycardia while claiming that she was off the antihypertensive medications. Wide fluctuations in blood pressure and heart rate may be found with autonomic dysfunction, pheo-

chromocytoma, and medications. Her urinary metanephrines were normal previously and she claimed not to be taking antihypertensive medications. Autonomic function as assessed by non-invasive cardiovascular reflexes and sympathetic skin response has been found to be abnormal in patients with syringomyelia. Although the patient did not have an increase in the size of her hydrosyrinx, postural tachycardia syndrome has been known to develop long after decompression surgery. Fludrocortisone and β -blockers have been shown to improve these symptoms.

The second turning point of the case was readmission with persistence of symptoms and lack of response to fludrocortisone. A doctor-patient relationship is based on trust and the principle that both are working towards a common goal of betterment of the patient's disease. This balance is disturbed when one of the parties has an ulterior motive. This is a difficult situation where many physicians may be misled. Medical evaluations may yield positive results in patients with factitious disorders because these patients use physical means to produce their symptoms. The clues that may alert the physician are subtle and include a prolonged history, multiple previous hospital admissions without a definite diagnosis or therapy, and travel from a different state for medical care. The physicians were misled into placing a permanent pacemaker.

The final twist in the case presentation was the recurrence of symptoms, the finding of a self made rash, and the half empty bottle of clonidine found in the possession of the patient. Factitious disorders describe presentations in which the individual intentionally produces or feigns physical or psychological symptoms to satisfy a psychological need to assume a sick role. This is in contrast to malingering, in which the motivation is obvious external gain (for example, disability benefits), and conversion disorder where the symptoms are not intentionally produced. The American Psychiatric Association's *Diagnostic and Statistical Manual*, 4th edition, DSM-IV) classifies Munchausen's syndrome as a subtype of factitious disorder. Baron von Munchausen was a Hanoverian cavalry officer who told greatly exaggerated tales of his adventures and his name, "Munchausen" came to symbolise dramatic unbelievable stories with grandiose lies.⁴ In 1951 the term Munchausen's syndrome was coined by Asher to describe a syndrome of frequent hospital admissions for false symptoms and signs. Mehta and Khan identified 58 cases of cardiac Munchausen's syndrome in the med-

Box 2: Causes of syncope associated with bradycardia

- Vasovagal syncope.
- Carotid sinus hypersensitivity.
- Situational syncope.
- Glossopharyngeal neuralgia.
- Bradyarrhythmias:
 - Sinoatrial node dysfunction.
 - Atrioventricular nodal block.
- Medications.

Box 3: Causes of syncope associated with normal heart rate or tachycardia

Postural hypotension

- Volume depletion.
- Physical deconditioning.
- Drugs (antihypertensive, antidepressant).
- Neurogenic:
 - Peripheral (diabetic, nutritional, amyloidosis).
 - Central (multisystem atrophy).
- Idiopathic.

Tachyarrhythmias

- Supraventricular tachycardia, atrial flutter/fibrillation, ventricular tachycardia/fibrillation.

Aortic outflow tract obstruction

- Aortic stenosis, hypertrophic obstructive cardiomyopathy.

Mitral valvular obstruction

- Atrial myxoma, mitral stenosis.

ical literature. Of these patients, three had the predominant complaint of syncope, of whom two received a permanent pacemaker placement.⁵ So although rare, this has been reported in the past.

This case serves as a reminder to critically appraise the history and to consider a psychiatric aetiology when things are just not the way they should be.

Follow up

Subsequently a family meeting and psychiatric consultation were called. The family was counselled to discard all the old medicine bottles that the patient had in her possession. The pacemaker was removed at a later date. However the patient has been admitted four more times since then with infection at the site where the pulse generator had been extracted. The bacterial culture from the wound site grew *Enterococcus faecalis* consistent with deliberate faecal contamination. Psychiatric consultation of the patient is ongoing.

Final diagnosis

Munchausen's syndrome.

References

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