"Doc, I'm getting strokes!"

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A 17-year-old girl presents complaining of attacks of numbness from the waist up. The attacks start with a tingling sensation in her hands, which proceeds to the face, and then to her entire upper body. Some attacks have caused her to lose the use of her arms.

She has had several attacks in the past year or two, but they have become more frequent and severe in the past month. She is somewhat symptomatic at present, although not as bad as earlier in the day. She denies a history of migraine, or a family history thereof, and says she has not felt stressed recently.

A detailed clinical exam was done, with special attention paid to her cardiac and nervous systems (Table 1).

Table 1
Exam results

- A subjective, symmetrical decrease in sensation upon light touch of her face
- Temperature: 36.2 C
- Pulse: 76 beats/minute
- Respiratory rate: 18 breaths/minute
- Blood pressure: 96/60 mmHg
- Electrocardiogram: Normal

Questions:

1. What is the differential diagnosis?
2. What is the most likely diagnosis, and what does it mean?
3. Can we confirm the diagnosis?
4. What is the best therapeutic approach?

Answers:

1. What is the differential diagnosis?

The differential diagnosis is broad, although her symptoms are not typical of a particular condition. Possibilities include atypical migraine, supraventricular tachycardia, hyperventilation secondary to acidosis or respiratory illness, somatoform disorder, or chronic hyperventilation syndrome (CHS).

2. What is the most likely diagnosis, and what does it mean?

CHS can be a disabling disorder which is frequently overlooked. CHS typically presents with multiple complaints without much supporting evidence of disease. The precise cause of the hyperventilation is uncertain, but it is believed to be due to an excessive response to endogenous respiratory stimulants, such as sodium lactate, or exogenous stimulants, such as stress, caffeine, or carbon dioxide (CO_2) exposure. Although generally benign from an emergency point of view, hyperventilation has been associated with syncope, seizures, and coronary vasospasm in patients with coronary occlusive disease.

In contrast to acute hyperventilation (AH), which is generally clinically obvious, CHS may be difficult to diagnose because hyperventilation is often inapparent, and patients tend to focus on complaints not immediately associated with the respiratory system. Careful observation will often reveal frequent deep sighing respirations (two to three per minute). Chronic respiratory alkalosis is often compensated by renal bicarbonate excretion, resulting in a normal pH despite persistently low CO_2. Additional respiratory stimulation, provoked by further stress may lead more rapidly to symptoms, as the acid-base buffering ability is reduced. This is likely the case in our patient, where the muscle stiffness is likely carpopedal spasm due to acute-on-chronic respiratory alkalosis.

Patients with CHS are usually overtly anxious, however, the condition may occur in the presence of apparent calmness. The incidence of hyperventilation syndrome is increased in first-degree relatives.

3. Can we confirm the diagnosis?

Symptoms of CHS commonly imitate one or more of a variety of other clinical syndromes. The commonest complaints are of lightheadedness, dizziness, numbness, dyspnea, or chest pain, although numerous other symptoms are often reported. Electrocardiogram changes may be present, resulting in further overlap between acute coronary syndrome or pulmonary embolism (PE). Gastrointestinal symptoms are also common. The key to making the diagnosis is to consider all the symptoms together, rather than trying to explain each symptom in isolation. A history of previous similar episodes is helpful in diagnosing CHS.

In newly suspected cases, dangerous conditions associated with hyperventilation, like metabolic acidosis, PE, or cardiac arrhythmias should be excluded. CHS should be diagnosed with considerable caution in older patients, or in those with a prior history of cardiac or respiratory disease. Normal pulse oximetry can occur in severe respiratory illness and, thus, should not be used alone to support the diagnosis. Arterial blood gas analysis in CHS will usually show a compensated respiratory alkalosis. For patients in whom symptoms have settled, reproduction of symptoms with intentional hyperventilation (30 breaths per minute for up to three to four minutes) will sometimes help clinch the diagnosis, and help convince patients they do not have a serious illness.

4. What is the best therapeutic approach?

The most important thing to do in the emergency department (ED) is to rule out life-threatening conditions associated with hyperventilation. Initial interventions, once the diagnosis is strongly suspected, include gentle reassurance with a clear description of what is happening.

Breathing expired air from a paper bag, as is frequently advocated for acute hyperventilation, is controversial. The technique could lead to hypoxia, especially in cases where CHS is misdiagnosed. In selected cases, breathing expired air from a paper bag may be indicated for exacerbations, in that it may provide symptomatic relief, empower patients, and help them understand the basic pathophysiology.

Further interventions include elective referral for instruction on optimal (diaphragmatic, as opposed to thoracic) breathing techniques, and stress reduction through relaxation therapy, counselling, or pharmacologic means.

More on our patient...

The patient's symptoms recur within 20 seconds of voluntary hyperventilation. When the nature of the illness is explained, the patient's mother points out that the patient is a perfectionist, and has been working extremely hard at school. The mother also admits taking medication for anxiety. In view of the chronic duration of the "attacks," more sinister conditions were felt to be unlikely enough for her to be discharged from the ED without further tests. Dx

This department covers selected points to avoid pitfalls and improve patient care by family physicians in the ED. Submissions and feedback can be sent to diagnosis@sta.ca.

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