REVIEW

Recognition of acute illness in people with chronic neurological disability

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Coincidental acute illness is common in patients with chronic progressive or stable neurological disability. Although the presentation of acute illness in these patients is often similar to that in people without pre-existing disability, sometimes it may be atypical. In addition, an adequate medical history is often difficult to obtain in patients with advanced neurological disease, for example because of the presence of aphasia, severe dysarthria, or cognitive impairment. In these circumstances early and accurate diagnosis of acute medical and surgical emergencies requires both clinical acumen and a high index of suspicion. The aim of this article is to describe the main atypical manifestations of acute illness in patients with chronic disabling neurological disorders.

THE NATURE OF THE DISABLING NEUROLOGICAL DISEASE

The underlying neurological disorder that caused the disability may have rare or obscure features that are normally associated with other acute medical and surgical disorders. The occurrence of these symptoms de novo in the course of a stable or slowly progressive neurological disability may be mistaken for a new, unrelated illness. Alternatively, the symptoms and signs of an acute illness may be attributed to the neurological condition. Therefore, awareness of these features often has an important diagnostic value. A useful diagnostic clue in these situations is that symptoms attributable to the underlying disease usually resolve or improve significantly with better control of the neurological disease in question. For example, proctalgia is a rare symptom of Parkinson's disease and usually responds to treatment with levodopa drugs or dopamine agonists.⁶ If the rectal pain is alleviated with increase of the antiparkinsonian drugs and there are no other clinical signs of bowel disease, it would be reasonable to attribute the proctalgia to the Parkinson's disease. The logical management strategy then is to monitor the patient's condition periodically rather than to subject them to unnecessary invasive investigations. Further examples of rare features of common neurological disease are the sudden occurrence in patients with chronic multiple sclerosis of trigeminal neuralgia that may be misdiagnosed as dental pain⁷ or the presentation with unexplained, severe electrolyte imbalance. (Symptomatic hyponatraemia attributable to the syndrome of inappropriate antidiuretic hormone secretion has been reported in patients with multiple sclerosis and was shown to be because of foci of inflammation in the supraoptic nuclei of the hypothalamus).^{8,9}

Diagnostic difficulties may also arise when the acute illness triggers a serious complication of the original neurological disorder. The dramatic nature of these complications may overshadow the symptoms of the acute illness. This scenario is particularly common in patients with acute or chronic high thoracic or cervical spinal cord injury. In a review of 1300 patients with chronic paraplegia all those who, during the study follow up period, had an acute abdomen attributable to a perforated gastric or duodenal ulcer, intestinal obstruction, or peritonitis presented with symptoms of autonomic dysreflexia (sudden, severe headache, profuse sweating, and severe hypertension). Other features included increase in the severity of muscle spasticity, shoulder tip pain, or vomiting. The classic signs of acute abdomen,

A comprehensive study of the prevalence of disability in the 1980s has shown that more than 6 million people in the United Kingdom had a chronic disability. Neurological disease accounted for 13% of all disabling conditions and was the cause of long term disability in about 2% of the whole British population.^{1,2} A decade later a large epidemiological survey³ reported an increase in the incidence and prevalence of neurological disability. It has also been suggested that this trend is likely to continue because of a longer life expectancy⁴ and the significant improvement in medical care in the acute and chronic phase of severe brain disease or injury that would have been fatal in the past.⁵ Medical practitioners in all specialties are therefore likely to see patients with chronic neurological disability. The classic symptoms and signs of acute illness may be absent in patients with chronic neurological disability and a better understanding of the atypical presentation of acute coincidental disease in these patients may help to avoid delays in the diagnosis and treatment of medical and surgical emergencies.

The difficulty in reaching the correct diagnosis of acute coincidental illness in patients with neurological disability may result from the inherent nature of the underlying chronic neurological disease itself or from its treatment. It is also often because of the difficulty in obtaining an adequate medical history. In some patients psychiatric morbidity, such as depression, or manipulative behaviour, adds to the difficulty of making an accurate diagnosis. Any or all of these factors may be present in the same patient.

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such as abdominal muscle rigidity (guarding), rebound tenderness, fever, and neutrophil leucocytosis, were often absent.¹⁰ The diagnosis of an acute abdomen can be easily missed in these circumstances unless, because of a high index of suspicion, the appropriate diagnostic investigations are carried out.

On the other hand, the symptoms of a *de novo* and unrelated disease may be attributed to the underlying chronic neurological disorder. For instance, it is not uncommon that the difficulties with swallowing attributable to oesophageal candida infection in a patient with multiple sclerosis, parkinsonism, or motor neurone disease are misdiagnosed as neurogenic dysphagia caused by progression of the underlying neurological disease.

THE EFFECT OF DRUG TREATMENT

Some of the drugs that are used to control the symptoms or modify the course of neurological diseases can change the manifestations of acute illness. Disease modifying drugs, such as corticosteroids or immunosuppressants, often mask the clinical features of systemic infection. On the other hand, the sudden withdrawal of some drugs, for example, muscle relaxants or neuroleptic drugs, may result in symptoms indistinguishable from those of severe infection. These symptoms include fever, tachycardia, hypotension, and mental confusion and may arise after the sudden withdrawal of treatment with baclofen,¹¹ phenothiazines, or anticholinergic drugs.¹²

THE MEDICAL HISTORY

The importance of the medical history for clinical diagnosis cannot be over-emphasised. A well taken medical history is usually sufficient for making the final clinical diagnosis, or at least a shortlist of differential diagnoses. Gruppen *et al*¹³ have found that just by analysing the patient's main complaints doctors were able to make the correct diagnosis in 78% of cases. Interestingly, the physical examination and laboratory tests were found to be mostly useful for excluding alternative diagnoses. An additional value of the medical history is that it improves the accuracy of the reporting and interpretation of laboratory tests and other investigations. This finding was illustrated well by a study that examined the effect of providing clinical information on the quality of reporting CT scans by experienced radiologists. The authors found that a panel of consultant radiologists correctly reported CT scans much more frequently when adequate information about the patient's clinical presentation was provided than when this information was withheld. Furthermore, the radiologists' diagnostic accuracy deteriorated when the provided clinical data were incorrect.¹⁴

Although some patients tend to underreport symptoms of cognitive impairment,¹⁵ most patients with chronic neurological disease are able to give an accurate description of their symptoms and medical history, even when they are frail or mentally ill.¹⁶ In most cases the clinician should be able to obtain an adequate medical history, even when aphasia or cognitive impairment is present. The patient's family members, professional caregivers, or general practitioner may also provide valuable diagnostic information. The importance of obtaining a history from the patient's carers should not be underestimated, as the recognition of acute illness in patients who are not able to report their symptoms frequently depends on their carers' ability to observe and report changes in the patient's functions or behaviour.

PSYCHIATRIC MORBIDITY

Psychiatric morbidity is common in patients with neurological disease. For example, in one study¹⁷ as many as 18% of patients presenting with traumatic brain injury were found to

Changes that suggest an acute illness in patients with chronic neurological disability

Unexplained deterioration in functional abilities

- Reduced mobility, falls
- New difficulties with performing activities of daily living
- Increased dependency on carers

Increase in the neurological impairment

- Increase in muscle spasticity and/ or muscle spasms
- Sudden reduction in muscle strength of a partially weak limb
- Worsening of dysphagia or dysarthria
- Increase in the frequency of epileptic seizures

Change in cognitive function or behaviour

- Acute mental confusion
- Sudden deterioration in pre-existing dementia
- Irritability, agitation, aggressive behaviour

Non-specific symptoms

- Sudden, significant changes in blood pressure and/ or heart rate
- Excessive fatigue, lethargy, sleepiness
- Urinary incontinence occurring *de novo*

have a history of major psychiatric illness. A wide range of disorders may occur, including depression, anxiety, and mania. Poor mental health is often associated with deterioration in functional capabilities¹⁸ that may be attributed to the primary neurological disease.

In addition to genuine psychiatric morbidity, simulation of physical symptoms, such as convulsions, visual field loss, etc, also seems to be common in patients with neurological disability. Eames¹⁹ reported that 54 of 167 patients with traumatic brain injury referred for treatment of severe behavioural disorders were found to have clinical features of gross hysteria. Malingering has also been reported in some patients. A good analysis of the patient's symptoms and a carefully conducted clinical examination may clinch the diagnosis, but in some cases admission to hospital for observation may be necessary. A classic example is when a patient with epilepsy is also suspected of having frequent pseudoseizures. In this situation, observation in hospital by experienced medical and nursing staff (possibly supplemented with telemetry) is invaluable.

THE ATYPICAL MANIFESTATIONS OF ACUTE ILLNESS

The atypical presentations of acute illness in patients with chronic neurological disability, summarised in the box, include the sudden or gradual deterioration in the patient's functional abilities, increase in the severity of a previously stable motor or cognitive impairment, change in behaviour, or subtle non-specific complaints.

Functional decompensation

Unexplained deterioration in mobility in a previously ambulant disabled person and/or increase in the dependency on others for assistance with personal care or other activities of daily living should alert the clinician to the possibility of an acute intercurrent illness. Respiratory and urinary tract infections are by far the commonest causes. Other causes include dehydration, electrolyte imbalance, drug adverse effects, and accidental falls.

Increase in the severity of the underlying neurological impairment

A sudden increase in the severity of spasticity, dysphagia, and dysarthria, or in the frequency of epileptic seizures may be the sole manifestation of an acute illness in a person with chronic neurological disease. Acute illness in patients with chronic upper motor neurone lesions frequently leads to the development of flexor or extensor muscle spasms or severe increase in a pre-existing hypertonia. Factors that trigger or aggravate these symptoms include bacterial infection, pressure sores, bladder stones, and anal fissures.²⁰ Systemic infection is also a potent cause of increased seizure frequency in patients with well controlled epilepsy. It may even precipitate status epilepticus. Acute infection was found to be the cause of status epilepticus in 50% of neurological patients presenting with this life threatening medical emergency.²¹

Behavioural change

Patients with global aphasia and those with severe cognitive impairment are usually not able to give account of their symptoms. Irritability, agitation and aggressive behaviour are common features of acute illness in this patient population, especially when pain is a major feature.

Non-specific symptoms

Acute changes, such as a sudden unexplained rise in blood pressure and heart rate, profuse sweating or agitation, are often attributable to severe discomfort or pain. These clinical features are particularly significant in patients who are unable to communicate their distress because of severe language impairment or reduced level of consciousness, for example, those in the persistent vegetative state. Joint and localised soft tissue pain attributable to complications of the underlying neurological disease is frequently misdiagnosed. For example, shoulder pain attributable to adhesive capsulitis occurs in 50% of hemiplegic stroke patients,²² but the correct diagnosis is often missed because the pain is usually attributed to the patient's pre-existing degenerative arthritis. Lethargy, sleepiness, falls, and acute mental confusion may also be the only manifestation of an acute intercurrent illness. Delay in the diagnosis of coincidental illness may occur because patients with neurological disease often attribute new symptoms or deterioration in pre-existing symptoms to fatigue and may not seek medical help.

CONCLUSION

Symptoms occurring de novo or deterioration in the patient's functional abilities in the course of a chronic neurological

disease should not be attributed to the underlying disorder until other intercurrent illness has been excluded.

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