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A 14-Year-Old Boy from Rural Tanzania With Difficulty in Walking

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Clinical Presentation

History

A 14-year-old boy presents to his local hospital in rural Tanzania with a history of difficulty in walking. He had been well until just over 2 years earlier when his illness suddenly started. He describes that he was walking home from school when he first noticed that his legs began to feel heavy and started to tremble, which caused him difficulty in walking, with a tendency to fall over. He slowly completed the journey home; and since that day, he has been unable to stand or walk unaided. He now stands on his toes and drags his legs around with the aid of a stick (Fig. 24.1). He denies any history of fever, pain, sensory, bladder or bowel symptoms or disease progression.

His history is otherwise unremarkable. He lives in a village in northern Tanzania in a rural area with limited agricultural suitability, where the main staple food crop is cassava. His diet for the 2 months before the illness was almost exclusively cassava. He has three other siblings, one of whom is similarly affected, whereas their parents remained fine. He mentions that identical cases had occurred in his own and neighbouring villages at around the same time.

Clinical Examination

Clinically, he is well nourished with normal vital signs. General examination is unremarkable. On neurological examination, he is fully orientated and higher mental function appears normal. Cranial nerves are normal but bilateral optic pallor was noted on fundoscopy. Limbs reveal signs of spastic paraparesis with flexion contractures at both ankles and knees. Power in the legs is graded 3 to 4 out of 5 (= just overcoming gravity), with the knee extensors and foot dorsiflexors involved to the greatest extent (Fig. 24.1). There is bilateral hypertonia, hyperreflexia and sustained ankle clonus with extensor plantar responses. The arms are normal apart from generalized hyperreflexia. There is no impairment or loss of sensation. A lumbar lordosis with thoracic kyphoscoliosis is noticeable only on standing.

• **Fig. 24.1** A 14-year-old boy from rural Tanzania with spastic paraparesis. His illness started about 2 years earlier and had an acute onset. Several other people are also affected in his own and neighbouring villages.

Investigations

Full blood count, erythrocyte sedimentation rate, blood glucose and creatinine are normal. Urine analysis is normal. Microscopy of urine and stool specimens does not show any ova of *Schistosoma* species. HIV serology and VDRL are negative. Lumbar puncture is normal. Radiographs of the chest and thoracolumbar spine are normal.

Questions

1. What is the clinical diagnosis and the likely cause in this patient?
2. How do you plan to manage this patient?

Discussion

A 14-year-old boy from rural northern Tanzania presents with acute-onset non-progressive spastic paraplegia. There is no history of back pain. On examination, there is no sensory impairment and no bladder dysfunction. The main staple food crop in his village is cassava.

Answer to Question 1

What is the Clinical Diagnosis?

The clinical syndrome is spastic paraparesis. The main differential diagnosis in Africa includes spinal tuberculosis (Pott's disease), transverse myelitis, spinal cord infections such as schistosomiasis and tuberculous myelitis, spinal malignancy (mainly metastases) and tropical nutritional myeloneuropathies.

There are three important features in our case: (1) the isolated involvement of motor neurons without any sensory and bladder involvement; (2) the absence of back pain; and (3) the acute onset with no progression over 2 years. These three clinical points make spinal tuberculosis, spinal cord infection or spinal malignancy very unlikely. Of note, his diet (and probably that of his siblings and other children in the village) for the 2 months before the illness was almost exclusively cassava, and the same disease has affected one of his siblings and more children in the neighborhood. Hence, a nutritional cause must be suspected.

The tropical myeloneuropathies that are nutritional in origin are konzo and lathyrism. Lathyrism in Africa occurs exclusively in Ethiopia. The clinical diagnosis in our patient is konzo. Konzo is a distinct form of tropical spastic paraparesis which occurs exclusively in cassava-growing areas in Africa.

Answer to Question 2

How Do You Plan to Manage this Patient?

There is no cure for patients with konzo because it results in a permanent spastic paraparesis. Management is therefore directed at support, symptomatic improvement and disease prevention. The majority of patients can walk with the aid of a stick or crutches and some will benefit from a wheelchair. Muscle relaxants have a limited role because of relative ineffectiveness as a result of the severity of the spasticity, and these agents' high long-term cost. Surgical treatment involving Achilles tendon lengthening operations have proved useful in improving mobilization in selected patients with konzo. Because of the severity of contractures in this patient, he should be assessed for surgery.

The Case Continued…

At follow-up examination at 6 and 12 months, the findings were unchanged, with a permanent spastic paraparesis. The patient had not been referred for surgery because of the lack of resources and the extent of the epidemic, with many other similar cases in the community. He uses one stick with very restricted mobility and works as a shoe repairer in his village.

SUMMARY BOX

Konzo

Konzo is a distinct form of tropical spastic paraparesis, which occurs exclusively in cassava-growing areas in Africa. The word "konzo" means "tied legs" in the language of the Yaka tribe in the DRC. It is characterized by an abrupt onset of a permanent but non-progressive form of spastic paraparesis related to cassava consumption. It occurs mainly as epidemics, typically during droughts, famines or armed conflict when there is an overreliance on cassava as a staple food for weeks or months. It also occurs in an endemic form but at much lower rates. It affects mainly children and breastfeeding mothers.

The following are the clinical criteria for diagnosing konzo:

- symmetrical spastic paraparesis without sensory or genitourinary involvement
- abrupt onset in less than 1 week with a non-progressive course
- occurring in a cassava-growing area, usually with other cases emerging at the same time
- no other cause found

Its cause is attributed to chronic high dietary exposure of cyanogenic glycosides from insufficiently processed cassava tubers, but the exact pathogenic mechanisms of konzo remain unknown.

Cassava is a staple food for more than 600 million people. It grows well in poor soils and is resistant to drought, plant diseases, insects and animal predators, but it contains cyanogenic glycosides, mainly linamarin. Processing disrupts the tubers releasing cyanide, which makes the food safe for human consumption. Konzo is associated with high intake of insufficiently processed cassava tubers in combination with low or absent levels of the essential amino acids methionine or cysteine. Oxidative stress and glutamate-mediated neuro-excitatory cell death appears likely to be the final pathogenic mechanism. This results in a clinically exclusive pattern of upper motor neurone disease.

Further Reading

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