Dystonia and choreo-athetosis in Wernicke’s encephalopathy

A case report

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Summary

An Indian man with Wernicke’s encephalopathy had nystagmus, pupillary changes and confusion, but the unusual and prominent features in his presentation were marked dystonia and choreo-athetosis, which responded rapidly to thiamine. The possible pathogenesis is discussed.

Severe Wernicke’s encephalopathy can present with coma, pupillary abnormalities, loss of oculocephalic and caloric responses, hypothermia and also partial complex seizures. However, to the best of our knowledge, dystonia and choreo-athetosis have not been previously described.

Case report

A 31-year-old Indian man presented to hospital with a 2-day history of persistent vomiting following a 2-week fast. He had complained of occasional dyspepsia for 1 year. There was no history of alcohol consumption. No anti-emetic agents such as metoclopramide or prochlorperazine were used either before or on admission to hospital. The patient had no past or family history of abnormal movements.

On examination the patient was dehydrated and hypotensive (blood pressure 90/60 mmHg). Positive findings were confined to the neurological system. He was conscious on admission but rapidly became confused. Horizontal jerk nystagmus was present with the fast component in the direction of gaze. The pupils were small and sluggishly responsive to light. Eye movements were full. Detailed cerebellar examination was not possible. The motor system, other cranial nerves, fundal examination and sensory testing were normal. He subsequently developed severe retrocollis and marked dystonic movements of his upper limbs, neck and facies. At times he exhibited groping and swaying movements of his upper limbs; bilateral choreo-athetosis was noted at other times. Coma ensued.

Laboratory investigations revealed: serum sodium 131 mmol/l; potassium 1,2 mmol/l; chloride 72 mmol/l; alkalai reserve 72 mmol/l; urea 17,9 mmol/l; and creatinine 203 mmol/l. The blood glucose level was 6 mmol/l; serum calcium 2,41 mmol/l (corrected); phosphate 1,45 mmol/l; amylase 191 mmol/l; bilirubin 27 mmol/l; magnesium 0,97 mmol/l; copper 16,7 μmol/l (normal 10,6-21 μmol/l); and caeruloplasmin 0,29 g/l (normal 0,12-0,45 g/l). A lumbar puncture and computed tomography of the brain were normal. Endoscopy showed gastric outlet obstruction as a result of duodenal ulceration.

Wernicke’s encephalopathy was diagnosed and the patient was treated with intravenous thiamine 100 mg immediately and hourly thereafter. He was also hydrated and given intravenous potassium. Within 4 hours all the dystonic movements disappeared and he recovered full consciousness. Over the next few days the electrolyte imbalance was corrected. The duodenal ulcer was treated conservatively with tri-K-di-citrato bismuthate and the patient remains well 6 months later.

Discussion

It is felt that our patient had Wernicke’s encephalopathy because of the presence of nystagmus, confusion, pupillary abnormalities and rapid lapse into coma. The quick response to thiamine also supports this diagnosis. However, tests for red blood cell transketolase and for thiamine pyrophosphate effect were not performed to prove the diagnosis. The cause of the deficiency was persistent vomiting consequent upon gastric outlet obstruction and fasting. The unusual feature in this patient was severe dystonia and choreo-athetosis. He had no history of abnormal movements; neither did he have a history of psychotropic drug ingestion. Copper metabolism was normal. Since the abnormal movements occurred together with other features of Wernicke’s encephalopathy and disappeared rapidly with thiamine administration, we feel that the dystonia was related to thiamine deficiency. Coma and pupillary changes, absent oculocephalic and caloric responses, hypothermia and even partial complex seizures have been reported in Wernicke’s encephalopathy. To the best of our knowledge, dystonia has not been previously documented previously.

Dystonia has been variously attributed to functional lesions of dopaminergic tracts of basal ganglia, to changes in striatal acetylcholine level, which can be modulated by dihydroxyphenylalanine or 5-hydroxytryptamine, or to structural defects in the brainstem loop modulated by striatal dopamine or increased acetylcholine levels centrally. Thiamine deficiency is known to interfere with the synthesis of neurotransmitter amino acids, e.g. glutamate and aspartate. High doses of glutamate, choline and 5-hydroxytryptamine have been successfully used in dystonic diseases.

Basal ganglia damage has been described in Wernicke’s encephalopathy. Thomson et al. described lesions in the basal ganglia, thalamic and subthalamic nuclei in animals with thiamine deficiency. However, Harper in an extensive study on 131 necropsies did not describe lesions in the basal ganglia, which were sectioned for histological examination. Torvik described lesions found at necropsy, especially in the thalamus, mammillary bodies and peri-aqueductal and periventricular areas. There is a gradual reduction of neurons in Wernicke’s encephalopathy compared with the sudden necrosis of axons. It is possible that a deficiency of selective neurotransmitter amino acids resulted in abnormal movements in this patient. Thiamine deficiency could also have affected the dopamine metabolism in these basal ganglia nuclei, resulting in an imbalance of acetylcholine or dopamine. This imbalance might have accounted for the dystonia and choreo-athetosis observed in this patient.

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Multiple lumbar spondylolyses with transverse process pseudo-arthroses

A case report

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Summary

An unusual case of multiple spondylolyses, spondylolisthesis at L5/S1, and pseudo-arthroses of the transverse processes of L4 is described.


Spondylolysis is a congenital or acquired defect in the pars interarticularis of a vertebral body and occurs throughout the spine, most commonly at the L4 and L5 levels. Multiple levels of spondylolysis are uncommon, but there have been 10 different reports and the condition seems more frequent in Eskimos, who are also more prone to spondylolysis in general.

Spondylolisthesis (anterior translation of a vertebral body in relation to the one below) occurs most frequently on the basis of spondylolysis or degenerative disease. Further causes and a classification are by Wiltse et al.

Case report

A 35-year-old farm labourer presented to hospital with a 7-year history of severe lower back pain radiating into the left lower leg. No history of precipitating events was obtained but the pain was aggravated by his work.

A mildly positive left straight-leg raising test with slightly reduced sensation and a questionable reduction in power in the left S1 distribution were noted.

Lumbar puncture revealed normal cerebrospinal fluid. Routine haematological and biochemical tests were normal.

A lateral radiograph of the lumbar spine revealed bilateral spondylolyses at L3 and L5 with a single spondylolysis at L4 and grade 2 spondylolisthesis at the L5/S1 level (Fig. 1). Pseudo-arthroses of both the transverse processes of L4 were seen (Fig. 2). The results of myelography were normal. Management was conservative.

Discussion

In the 18 reported cases of multiple spondylolysis of at least three levels, the spondylolyses were most frequently bilateral.