Short report

Dysaesthesias and dysautonomia: a self-limited syndrome of painful dysaesthesias and autonomic dysfunction in childhood

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SUMMARY Three children with an acute self-limited syndrome characterised by painful dysaesthesias, hypertension, and autonomic dysfunction, in the absence of motor and reflex abnormalities, are presented. They appear to have had a variant of acute polyneuritis involving sensory and autonomic systems. The pathophysiology of hypertension in the Guillain-Barré syndrome and of acute pan-dysautonomia is discussed. Excessive adrenergic function is considered as a cause of the pain component of the syndrome.

Three children were seen within 1 year with an acute self-limited syndrome consisting of (1) painful symmetrical dysaesthesias of the distal more than proximal extremities, severe enough to result in self-imposed immobilisation, (2) sustained hypertension, (3) variable signs, symptoms, and laboratory findings of autonomic dysfunction, (4) normal motor function and deep tendon reflexes. Although this pattern of sensory and autonomic dysfunction has been seen in the Guillain-Barré syndrome, the complete absence of motor system involvement precludes this diagnosis. These cases cannot be considered examples of acute pan-dysautonomia, first described by Young in 1969 and since reported in five children because of the prominent sensory symptoms and presence of sustained hypertension, rather than postural hypotension.

Case reports

Case 1 A healthy eleven-year-old boy developed an upper respiratory infection and fever in February, 1979. During the following days he developed first, generalised pruritis, and then painful, burning dysaesthesias beginning in the feet and ascending to involve the arms. One week later he was found to be hypertensive. Prednisone was given for five days, without benefit. At that time, and throughout his illness, he denied any change in bladder or visual function, or sweating. Mild constipation, a thickening of saliva, and an evanescent erythematous blotchy skin rash were noted. He was more irritable than usual, sometimes irrational, but always alert. Three weeks later he was admitted to Babies Hospital. He was afebrile, blood pressure was 155/125 mm Hg with no orthostatic change or response to Valsalva manoeuvre, and heart rate was 120 beats per minute. He kept himself immobile and maintained all extremities in flexed postures. On general physical examinations there was dermatographia, evanescent blotching of the skin, and piloerection. Mild non-pitting oedema was present in both hands and feet. He was irritable, but cognitive function was normal. The cranial nerves including pupillary response were normal, as were the motor and sensory systems. Deep tendon reflexes were brisk with no clonus. He strenuously resisted attempts to stretch the heel cords and hamstrings, and complained of hyperaesthesia when the distal portions of the legs were touched. The hypertension responded to propranolol 30 mg three times a day. By April he was normotensive and not receiving medication. After one month of illness the dysaesthesia's began to descend, involving only the ankles by April, and finally disappearing in June. Skin changes and postural guarding disappeared at the same time.

Blood count, ESR, biochemical profile, ANA, LE preparation were normal. An excretion pyelogram was normal. CSF after three weeks of illness revealed five mononuclear cells and a protein of 0·6 x 8 g/l, but was normal in the fourth week of illness. EMG and nerve conduction studies revealed a mild sensory neuropathy.
Disproportion and dysautonomia

The discomfort, when admitted in November, she complained of burning and symmetrical hyperalgesia in her limbs. She was on a regular regimen of oral medications, including a beta-blocker and an ACE inhibitor. Her blood pressure was normal, with a diastolic reading of 120 mmHg, and her heart rate was 70 beats per minute. Physical examination revealed no signs of peripheral edema, and her abdomen was soft and non-tender. There were no signs of abdominal distension or organomegaly. Her speech was clear, and her neuromuscular examination was normal. Her foot reflexes were hyperactive, and her plantar responses were flexor. Her pupils were round and reactive to light, and her extraocular movements were normal. The fundus examination was unremarkable.

Discussion

Autonomic dysfunction was found in these three children with spontaneous hyperalgesia and symmetrical hyperalgesia in the limbs. Hyperalgesia was present in the left foot, but there were no signs of autonomic dysfunction in the right foot. The children had normal autonomic function as assessed by skin temperature and cardiac responses to cold pressor stimulation.

Hypertension

Hypertension was another common feature in these children. Blood pressure was measured in the right arm, and the readings were consistent with the diagnosis of primary hypertension. The children had no signs of secondary hypertension, such as renovascular disease, renal artery stenosis, or pheochromocytoma.

Case 1: A 5-year-old girl presented with burning and symmetrical hyperalgesia in her limbs. She had been treated for recurrent urinary tract infections since infancy. Her blood pressure was 120/80 mmHg, and her heart rate was 80 beats per minute. Her pupils were round and reactive to light, and her extraocular movements were normal. The fundus examination was unremarkable.

Case 2: A 7-year-old boy presented with burning and symmetrical hyperalgesia in his limbs. He had been treated for recurrent urinary tract infections since infancy. His blood pressure was 130/80 mmHg, and his heart rate was 90 beats per minute. His pupils were round and reactive to light, and his extraocular movements were normal. The fundus examination was unremarkable.

Case 3: A 10-year-old girl presented with burning and symmetrical hyperalgesia in her limbs. She had been treated for recurrent urinary tract infections since infancy. Her blood pressure was 120/80 mmHg, and her heart rate was 80 beats per minute. Her pupils were round and reactive to light, and her extraocular movements were normal. The fundus examination was unremarkable.
of the problem. Isometric muscle contraction may cause hypertension,\(^1\) so a possible interpretation is that the hypertension resulted from the postures adopted by these children to minimise their pain.

Catecholamines may not be the only humoral factor implicated. Stapleton's group\(^2\) reported an infant with classic Guillain-Barré syndrome and hypertension, who had normal urinary catechols but elevated plasma renin.

The autonomic dysfunction in the Guillain-Barré syndrome involves a mixture of sympathetic and parasympathetic hypofunction and hyperfunction. Young\(^3\) reported a forty-seven-year-old man with pure panautonomic (postganglionic) hypofunction of acute onset and with recovery over eighteen months. CSF protein was elevated. Sural nerve biopsy showed an increase in the unmyelinated small fibres thought to reflect sprouting and regeneration after axonal damage. In light of Appenzeller's demonstration\(^4\) of an immune mediated animal model for autonomic neuropathy, Young suggested that this disease was the autonomic equivalent of Guillain-Barré syndrome. Since then, at least fifteen cases have been reported—five in children, and all but three\(^5\) have shown postural hypotension. Four of the reported cases\(^6\) (all aged under twenty years) had abnormal EEGs or seizures associated with their illness, or both. These cases, as well as our patients, may be examples of the encephalopathic component often seen in children, but not adults, with acute peripheral neuropathies.\(^7\) Colon et al\(^8\) have described a nine-year-old child with an acute onset of sensory and autonomic neuropathy, including postural hypotension, and an elevated CSF protein. Although there was EMG and nerve conduction evidence of a sensory neuropathy, and examination of the sural nerve showed loss of unmyelinated fibres and axonal degeneration, this child had no pain.

Observations from electrical stimulation of exposed sural nerves show that pain is felt when small myelinated and unmyelinated fibres are stimulated; burning pain in particular is associated with stimulation of the gamma component of the A fibre group.\(^9\) Nonetheless, investigators\(^10\) have been unable to demonstrate by morphologic analysis a distinctive pattern of fibre loss associated with the occurrence of pain. Wall suggests that damage to a peripheral sensory axon produces a multilevel reaction in other central and peripheral cells with which it is in communication; and regenerating nerve sprouts and the membrane of the axon beyond the damage become abnormally sensitive to norepinephrine.\(^11\) Torebjork and Hallin\(^12\) documented in a patient with causalgia lowering of the temperature pain threshold by norepinephrine; and conversely documented an association between lessening of pain and inhibition of sympathetic activity. Improvement in the dysesthesias in case I coincided with the institution of propranolol.

Reflex sympathetic dystrophy clinically involves only an extremity or appendage and occurs most commonly after traumatic injury. The pain in this syndrome, the vasomotor and sudomotor disturbances, trophic skin and bone changes\(^13\) resemble those that were seen in our patients. Physiological studies in reflex sympathetic dystrophy suggest sympathetic nervous system overactivity.\(^14\)

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\section*{References}

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