

Cronkhite-Canada syndrome associated with peripheral neuropathy

The Cronkhite-Canada syndrome (CCS)^{1,2} is a rare idiopathic acquired generalised gastrointestinal polyposis syndrome associated with onychodystrophy, alopecia and skin hyperpigmentation. The associated protein-losing enteropathy and progressive malnutrition usually result in death. Here we describe a patient with CCS and peripheral neuropathy, a previously unreported association.

A 70-year-old male presented with a six month history of weight loss, diarrhoea, dystrophic nails, skin hyperpigmentation, numbness of the extremities and muscle weakness. His medications were digoxin, allopurinol, sublingual nitroglycerin and temazepam. Clinical examination revealed alopecia, generalised skin pigmentation, onychodystrophy, leukonychia, pedal oedema and atrophic skin over the feet. There was symmetrical proximal and distal muscle wasting and weakness in the upper and lower limbs (Medical Research Council grade 4/5). The upper limb reflexes were normal but the knee jerks were depressed and the ankle jerks were absent. Pain, temperature and light touch sensation were symmetrically reduced in a glove and stocking pattern distal to the wrists and mid-calves. Joint position and vibration sense were normal. His stance and gait were normal. Endoscopic and radiological studies revealed generalised intestinal polyposis. Histological examination of the polyps showed cystic dilatation of the glandular elements and chronic inflammation of the lamina propria consistent with a diagnosis of Cronkhite-Canada syndrome. A sural nerve biopsy showed a reduction in the number of large diameter myelinated fibres but no other specific features. Electromyography of quadriceps and extensor forearm muscles was normal. Nerve conduction studies of the right upper and lower limbs showed an absent peroneal nerve response, prolonged sensory action potential latencies and reduced sensory nerve action potential amplitudes. Because of his protein-losing enteropathy, nutritional causes of neuropathy were considered. He was extensively investigated for recognised nutritional deficiencies which cause neuropathy but no cause was identified. The patient was commenced on a high protein high calorie diet with oral vitamin supplementation. Four months after discharge he had lost a further 10 kg and he had become weaker although his sensory signs were unchanged. He was now unable to rise from a chair or climb a step. He died five months after diagnosis. An autopsy confirmed the presence of extensive polyposis extending from the stomach to the rectum. There were no adenomatous foci in the polyps examined and there was no macroscopic or microscopic evidence of carcinoma. Post mortem examination of the nervous system was not performed. The immediate cause of death was pulmonary embolism.

The late onset of weight loss, diarrhoea, onychodystrophy, generalised hyperpigmentation and alopecia and the finding of widespread gastrointestinal polyposis with cystic dilatation of the glandular elements and chronic inflammation of the lamina propria in a patient without a family history of polyposis established the diagnosis of CCS. The patient also had symptoms and signs of a symmetrical sensorimotor neuropathy. Sural nerve biopsy did not demonstrate any specific features. Electrophysiological studies demonstrated the presence of a peripheral neuropathy. This is the first report of the occurrence of peripheral neuropathy in CCS. Although patients with CCS and sensory symptoms have been previously reported^{1,3,4} none has been observed to have a peripheral neuropathy. Although a nutritional deficiency secondary to the gastrointestinal disease seems a possible explanation for this neuropathy we found no evidence to support this. We suggest that the neuropathy may be due to the disease process underlying the Cronkhite-Canada syndrome. ■

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