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Short report

A chronic and painless form of idiopathic brachial plexus neuropathy

GD SCHOTT

From the National Hospital for Nervous Diseases, Queen Square, London

SUMMARY Three patients are reported who developed slow and painless weakness and wasting of the muscles of one shoulder girdle, denervation in the affected muscles, and subsequent complete recovery. In two patients, an area of cutaneous sensory loss in the distribution of the circumflex nerve was detected. No underlying cause was found, and it is suggested that these features represent an unusual chronic and painless variant of idiopathic brachial plexus neuropathy.

The typical features of idiopathic brachial plexus neuropathy are the sudden onset of severe pain around the shoulder girdle and the subsequent subacute development of weakness and wasting of the periscapular muscles.[1] In this condition, often termed neuralgic amyotrophy, bilateral involvement may occur, and involvement of other muscle groups including those of the arm, pelvic girdle and leg and diaphragm is also recognised, as are familial cases. There may be areas of cutaneous sensory loss, especially in the territory supplied by the circumflex nerve when the shoulder girdle is involved. The prognosis is generally good, although recovery may be protracted; 80% of patients may be expected to recover within two years and nearly 90% by the end of three years.[2] If these typical features are absent, alternative diagnoses require consideration, in particular disorders of anterior horn cells and compressive lesions of the nerve roots and the brachial plexus.

The present report describes three patients who developed localised weakness and atrophy of muscles around one shoulder girdle, but in whom pain was entirely absent throughout the course of the illness and in whom the weakness and wasting developed very insidiously; complete recovery occurred within several months. It is suggested that such patients, who may give rise to concern about the possibility of more serious conditions, represent an atypical form of idiopathic brachial plexus neuropathy.

Address for reprint requests: Dr GD Schott, The National Hospital for Nervous Diseases, Queen Square, London, WC1N 3BG, UK.

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Case reports

Case 1 This 14-year-old schoolgirl noticed the gradual onset over about nine months of progressive inability to elevate her right arm. She found swimming very difficult and also putting her arm into a sleeve. She had noticed her right shoulder had dropped. Her left arm and lower limbs were normal, there was neither sensory nor sphincter disturbance and she had had no pain. She had not had any intercurrent infections, surgery or other trauma, and had not received any drugs or vaccinations. Examination revealed depression of the right shoulder which sloped forward, prominence of the right scapula but without any true winging, and wasting of the spinati. There was severe weakness of the right trapezius and deltoid muscles and moderate weakness of the spinati and rhomboids. Fasciculation was not seen. No abnormality was detected on sensory testing, the reflexes were normal, plantar responses were flexor, and examination of the neck and supraclavicular fossa was also normal. Investigations included a normal blood count, ESR, plasma electrolytes and urea, CSF, cervical spine radiograph and myelogram. Serological tests for syphilis were negative. EMG studies showed features of denervation in the right deltoid and trapezius muscles with normal motor and sensory nerve conduction studies in the right ulnar and median nerves. At review four months after her investigations, she reported her strength had almost returned to normal and examination confirmed the virtual disappearance of the abnormal signs.

Case 2 This 34-year-old mechanic had become aware of a depression in the muscles behind the left shoulder six months before coming to medical attention. This realisation occurred two weeks after an uneventful operation for pilonidal sinus. A month after this operation, he experi-

enced difficulty in elevating his left arm when decorating his house, and this was followed two weeks later by weakness when holding a newspaper outstretched in front of him. He also had difficulty in taking his arm out of a sleeve and in using the gear-lever when driving. Examination revealed severe wasting and weakness of the left spinati with moderate weakness of the deltoid, triceps, biceps and brachioradialis muscles. There was a patch of impaired cutaneous sensory loss over the lateral aspect of the left upper arm. The reflexes were normal, plantar responses were flexor and the cervical spine and supraclavicular fossa were also normal. Investigations included a normal blood picture, ESR, creatine kinase, serological tests for syphilis, and chest, cervical and thoracic spine radiographs; EMG studies showed evidence of denervation in the left infraspinatus muscle where profuse fibrillation potentials were seen. When reviewed three months later, there was considerable improvement but wasting of the spinati, especially infraspinatus, remained marked; there were no sensory changes. After a further three months, recovery was complete and there were no abnormal signs.

Case 3 This 15-year-old schoolgirl noticed over a period of two months that she had progressive difficulty in elevating the left arm. She had been unable to reach high objects and had difficulty in putting her arm in the sleeve of her coat and jackets. Neither pain nor sensory symptoms had occurred. On examination there was pronounced wasting and weakness of the spinati and deltoid muscles on the left, the deltoid being particularly severely involved. There was no fasciculation. There was questionable weakness of the left triceps and small muscles of the hand. A small patch of impaired cutaneous sensory loss was detected over the lateral aspect of the left upper arm, and tendon reflexes were absent in the left arm with preservation of reflexes in the other limbs and flexor plantar responses. Her spine was normal. Investigations showed a normal blood picture, ESR, blood sugar, Paul Bunnell screening test, creatine kinase, CSF, radiographs of the chest, cervical spine and shoulders. EMG studies showed features of denervation in the left deltoid muscle with positive sharp waves, a markedly reduced interference pattern with units up to 4 millivolts and an excess of polyphasic units. Similar but less marked changes were seen in the left brachioradialis and triceps muscles; a decreased interference pattern with single units firing at high rates was seen in the first dorsal interosseous muscle, extensor carpi radialis and extensor digitorum communis muscles; sensory and motor nerve conduction studies in the left median and ulnar nerves were normal. When reviewed two months later, her strength had returned to normal, all her reflexes could now be elicited and there were no abnormal signs.

Discussion

The concept of idiopathic brachial plexus neuropathy[1] (also described by other terms such as neuralgic amyotrophy,[3] cryptogenic brachial plexus neuropathy, localised neuritis of the shoulder girdle, and acute shoulder neuritis) first developed in the 1940's, when a number of authors described the syndrome of severe pain affecting the shoulder region, the rapid development of weakness and wasting of the shoulder girdle muscles and subsequent recovery in most cases. Fasciculation was sometimes noted in affected muscles and sensory impairment not infrequently occurred, although it tended to be comparatively unobtrusive. A number of reports have subsequently been published.[1] The pattern of the disease in individual patients has suggested involvement of single or multiple nerve roots, trunks or peripheral nerves, and there is indirect evidence that discrete nerve fibre bundle involvement in the brachial plexus may occur.[4] Neurophysiological studies may show evidence of axonal damage and demyelination; denervation may be seen in affected and also in clinically unaffected muscles. Although commonly occurring as an unheralded, acute illness, an identical condition may follow injections and innoculations, antecedent infections and trauma (as in Case 2). Despite the possibility that an immune process may underlie the condition, there are no specific investigations which help to elucidate what is a clinical diagnosis. The sedimentation rate may be slightly increased but immunological abnormalities have been found in neither blood nor CSF; the CSF is usually normal although occasionally a slightly raised protein content has been reported. Lack of more specific information from investigations, the variability of the clinical features, the good prognosis and absence of modern pathological material all conspire to make the nature and aetiology of the disorder obscure.

The present patients illustrate that a similar condition may arise, but with two striking differences from the classical picture: the complete absence of pain and the very slow development of muscle weakness and wasting. Indeed, in Case 2 the first symptom of muscular wasting was discovered by chance and ignored. Although it could be argued that these two features preclude the diagnosis of brachial plexus neuropathy, it seems more useful to suggest that a variant of the typical condition may occur: the selective involvement of the shoulder girdle muscles, the minor sensory impairment in the territory supplied by the circumflex nerve noted in two cases, the complete recovery over some months, and evidence of denervation in affected muscles are all consistent with the diagnosis. At no time were there features suggesting an inflammatory cause, neither was there evidence of a compressive condition such as cervical disc protrusion. Poliomyelitis and motor neuron disease require consideration in a patient without sensory impairment. Motor neuron disease in particular initially caused concern in Case 1, but the complete recovery that subsequently occurred excluded even the "benign focal amyotro-

phy" described by Engel.[5]

Pain occurs in nearly all cases of idiopathic brachial plexus neuropathy; it occurred before weakness developed in 97 of 99 patients reported by Tsairis *et al*,[2] and whether pain occurred in the remaining two is not stated. Although occasional cases without pain have been referred to in earlier reports,[3 6 7] little attention has been paid to this and the rapid development of weakness has allowed a diagnosis of brachial plexus neuropathy to be made with confidence. Fortunately the pain which may be very severe usually subsides in days or a few weeks, rarely continuing intermittently for a year; its mechanism is obscure. There is no specific treatment for either the typical pain or the muscle weakness; although steroids are said to reduce the pain, their use would clearly be inappropriate in this chronic painless variant.

The importance of recognising the atypical features of this benign disease lies in the fact that more major investigations such as myelography may be held in abeyance providing the patient remains under careful review. With further experience, it may be possible to give a more reassuring prognosis at an earlier stage.

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