Neuralgic amyotrophy as a presenting feature of infective endocarditis

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Abstract

A 35 year old man presented to his general practitioner with severe right shoulder pain and subsequent weakness and wasting of the muscles in the affected shoulder girdle three weeks after a dental filling. His symptoms persisted despite standard treatment. He developed malaise, night sweats, weight loss, a petechial rash and a microcytic anaemia. On admission to hospital three months after the start of his symptoms he had also developed splenomegaly and the murmur of aortic regurgitation. Investigations confirmed the diagnoses of infective endocarditis and neuralgic amyotrophy. In this case neuralgic amyotrophy appears to have been the presenting feature of infective endocarditis. This association has not previously been described.

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Neuralgic amyotrophy (also known as Parsonage-Turner syndrome, brachial neuritis, serum neuritis) is a condition which is generally regarded as being associated with viral infection or immunisation, though commonly no antecedent event is identified. It is very rarely diagnosed correctly at first because the severity of the pain leads to consideration of other causes,¹ and examination for winging of the scapula is often neglected. It does have a host of rarely reported associations though these have often been identified prior to the onset of this condition. We describe a case where neuralgic amyotrophy was the presenting feature of infective endocarditis and may have led to a delay in diagnosis. Despite this the patient made a good recovery after a prolonged course of intravenous antibiotics and valve replacement.

Case report

A 35 year old right handed window cleaner was referred urgently to the rheumatology outpatient department with a three month history of severe right shoulder pain, muscle wasting, and weakness in the affected shoulder girdle and arthralgia in his right elbow and wrist. Rheumatoid factor was positive at a titre of 1:640. His general practitioner had tried standard analgesic preparations, non-steroidal anti-inflammatory drugs, and injection of the joint with cortisone all with little effect. Non-steroidal anti-inflammatory agents had been stopped when the patient developed a microcytic anaemia. The appointment was

expedited further when the patient developed arthralgia in both knees, malaise, night sweats, decreased exercise tolerance, weight loss of 2 stones, and a petechial rash over the dorsum of both feet.

Further detailed history revealed he had had a dental filling three weeks before the onset of symptoms, no other problems with his oral health were identified at this time. Examination revealed multiple splinter haemorrhages and pulp infarcts in both hands, a purpuric rash on the dorsum of both feet, and a macular rash over the entire trunk. Pulse was regular at 80 beats/min and collapsing in nature. Blood pressure was 140/60 mm Hg and heart sounds were normal. The chest was clear. There was smooth, tender splenomegaly. Urine dipstick was positive to blood and protein. All movements of the right shoulder were painful. There was marked wasting of the deltoid, triceps, supraspinatus, and serratus anterior muscles with ill defined areas of sensory loss over the right forearm and winging of the right scapula (see fig 1). Polyarteritis nodosa was suspected clinically and the patient was admitted to hospital for further investigation a week later.

On admission examination was unchanged except for a short mid-systolic and long early diastolic murmur at the left sternal edge. At

Figure 1 Photographs showing wasting of the right shoulder girdle and winging of the right scapula. Due to active denervation, wasting is an early and often prominent feature of neuralgic amyotrophy. Winging of the scapula is caused by involvement of the long thoracic nerve (reproduced with patient's permission).

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at this point the clinical diagnoses of infective endocarditis and neuralgic amyotrophy were made.

Blood tests revealed a microcytic anaemia (haemoglobin 89 g/l, mean corpuscular volume 72.5 fl, white blood cells 5.6×10^9 /l, platelet count 243×10^9 /l), raised viscosity (1.84 cP), raised C reactive protein (75 mg/l), and mild renal impairment (urea 15.2 mmol/l, creatinine 219 μ mol/l). Serum iron was low at 5 μ mol/l with normal transferrin (2.04 g/l) and ferritin (189 μ g/l). Urine dipstick was positive for blood. Blood cultures grew *Streptococcus maltans* in seven out of eight bottles and both transoesophageal and transthoracic echocardiography showed a bicuspid aortic valve with eccentric moderate regurgitation and a mobile vegetation at its base. Shoulder and chest radiographs were normal. Nerve conduction studies demonstrated slight abnormalities in sensory conduction over the medial aspect of the right fore and upper arm with active denervation of the deltoid, biceps, triceps, brachioradialis, and flexor carpi ulnaris compatible with neuralgic amyotrophy.

Intravenous benzylpenicillin and netilmicin were started with rapid resolution of both rashes, normalisation of plasma viscosity and C reactive protein, and improvement of renal function. Anaemia remained constant but investigations for blood loss and haemolysis proved negative. Pain in the shoulder abated over the first week but wasting and weakness progressed. Over the six weeks of antibiotic treatment the left ventricle began to dilate so urgent aortic valve replacement was undertaken and successfully completed with resolution of all abnormal haematological and biochemical parameters. Shoulder wasting and weakness persists but is improving slowly with outpatient physiotherapy.

Discussion

Neuralgic amyotrophy typically presents with sudden onset of severe unilateral shoulder girdle pain followed by weakness and wasting of some of the C5, C6, and C7 innervated muscles. The long thoracic (nerve roots: C5 predominantly, C6, C7; supplies serratus anterior) and suprascapular nerves (nerve roots: C5; supplies supraspinatus and infraspinatus muscles) are the most commonly affected and involvement of serratus anterior with scapula winging is one of the most consistent signs.¹ Involvement may be patchy and include the trapezius, sternomastoid, and diaphragm. The right arm is more frequently involved than the left and bilateral involvement is reported to occur in up to one third of patients. There is a male preponderance of between 2 and 11.5:1.^{2,3} Diagnosis is clinical, investigations mainly aiding in the exclusion of alternative diagnoses. No treatment is known to affect the long term outlook but steroids may diminish the severity of pain in some cases and full symptomatic recovery of strength occurs in 90% by three years.^{2,3} Hereditary forms of the disorder (hereditary neuralgic amyotrophy, McKusick number 162100 and hereditary neuropathy with liability to pressure palsies, McKu-

Learning/summary points

- Neuralgic amyotrophy should be suspected in patients presenting with severe shoulder/limb pain and weakness and winging of the scapula should be looked for in this context
- The associations of neuralgic amyotrophy should be considered at presentation and antecedent illness looked for. It should be remembered that most commonly the associations are benign and full recovery occurs in 90%
- Endocarditis should now be considered as one of the associations and signs of this condition looked for at diagnosis

sick number 162500⁴) exist but in sporadic cases the cause is unknown. Both cellular and humoral immune mechanisms have been postulated⁵ and there are myriad associations. The most commonly reported of these are serum sickness and inoculations. Others include pregnancy, viral infections (Epstein-Barr virus, cytomegalovirus, parvovirus B19,⁶ HIV seropositivity⁷), malignancy, post-streptokinase,⁸ paraproteinaemia,⁹ borrelia infections, brucella, IL2/interferon therapy, and the postoperative period.¹⁰ It has not, to our knowledge, been previously reported in association with infective endocarditis. Its temporal association in this case, its hypothesised immune aetiology, and the multiple immunological phenomena associated with endocarditis make infective endocarditis a most plausible cause.

Infective endocarditis is a serious condition associated with multiple immunological phenomena. Prompt treatment may obviate the need for valve replacement and decreases both morbidity and mortality. In this patient's case the diagnosis may have been delayed because the association with neuralgic amyotrophy was not recognised. We suggest that infective endocarditis be added to the list of associations for neuralgic amyotrophy.

- 1 Patten J. *Neurological differential diagnosis*. 2nd Ed. Springer Verlag, 1996: 290–1.
- 2 Tsairis P, Dyck PJ, Mulder DW. The natural history of brachial plexus neuropathy. *Arch Neurol* 1972;27:109–17.
- 3 Magee KR, DeJong RN. Paralytic brachial neuritis. *JAMA* 1960;174:1258–62.
- 4 Chance PFLM, Lipe H, Brown RH Sr, et al. Hereditary neuralgic amyotrophy and hereditary neuropathy with liability to pressure palsies: two distinct genetic disorders. *Neurology* 1994;44:2253–7.
- 5 Suarez GA, Giannini C, Bosch EP. Immune brachial plexus neuropathy: suggestive evidence for an inflammatory-immune pathogenesis. *Neurology* 1996;46:559–62.
- 6 Pellas F, Olivares JP, Xandoti C, et al. Neuralgic amyotrophy after parvovirus B19 infection (letter). *Lancet* 1993;342: 503–4.
- 7 Cordonnier C, Chazeraï P, Campagne JP, et al. Parsonage-Turner syndrome disclosing HIV seropositivity (letter). *Annales De Medecine Interne* 1993;144:346–7.
- 8 Fink GR, Haupt WF. Neuralgic amyotrophy following streptokinase thrombolytic therapy. *Deutsche Medizinische Wochenschrift* 1995;120:959–62.
- 9 Martinelli P, Macri S, Scaglione C, et al. Acute brachial plexus neuropathy as a presenting sign of peripheral nervous system involvement in paraproteinaemia (letter). *Acta Neurol Scand* 1997;95:319–20.
- 10 Griffin AC, Wood WG. Brachial plexitis: a rare and often misdiagnosed postoperative complication. *Aesthetic Plast Surg* 1996;20:263–5.