Isolated facial diplegia: a diagnostic dilemma

Introduction

Facial diplegia (bilateral peripheral facial palsy) is a rare clinical entity and occurs in 0.3–2% of facial paralysis patients (Stahl and Ferit, 1989). It can be a presenting feature of many diseases and presents a diagnostic dilemma to the physician. Facial diplegia merits further investigation since it could represent a serious underlying systemic disorder.

Discussion

Facial diplegia can have many causes (*Table 2*). Rarely there have been case reports of isolated facial diplegia in pregnancy (Kovacic et al, 1998), during human immunodeficiency virus (HIV) seroconversion (Wechsler and Ho, 2003) and following Hymenoptera sting (Raucq and Dupuis, 1998).

Lyme disease, caused by *Borrelia burgdorferi*, is the most common infectious cause of facial diplegia as shown by Clark et al (1985), and has become the most common vector-borne disease in Europe and North America (Barbour, 1998). Over 300 laboratory-confirmed cases are reported annually to the UK's Health Protection Agency. The mean annual incidence of Lyme disease in the UK is currently 0.64 cases per 100 000 total population.

It is important for emergency departments to recognize Lyme disease since it is a treatable condition. It is a multisystem and multistage infection which starts as an expanding red rash (erythema migrans) at the site of tick bite (stage 1). However, almost 25% patients do not exhibit this characteristic rash (*Figure 1*). Stage 2 (within several days to weeks) represents disseminated infection to skin, nervous system, heart and the joints manifesting as non-specific symptoms including malaise, fatigue, generalized myalgia and headaches.

Clark et al (1985) observed that cranial neuritis (including facial diplegia) frequently occurs alone and may be the presenting manifestation of the disease. In Europe and Asia, the first neurological sign is characteristically radicular pain followed by CSF pleocytosis (Bannwarth's syndrome) as observed by Steere (2001). After months to years (stage 3) persistent infection in the form of chronic arthritis of large joints, chronic encephalomyelitis and achrodermatitis atrophica chronicans may supervene. All stages are treatable with antibiotic therapy.

The diagnosis of Lyme disease is based primarily on clinical suspicion. The US Centers for Disease Control and Prevention (1999) developed a case definition of Lyme disease for surveillance purposes that includes either physician-diagnosed erythema migrans along with solitary lesions with diameters of at least 5 cm or at least one late joint, neurological or cardiac manifestation along with laboratory confirmation.

Serological confirmation is based on a two-test approach in which samples are first tested by enzyme-linked immunosorbant assay (ELISA) and those with equivocal or positive test are confirmed by western blotting. After the first 4–6 weeks of infection, 90% or more of patients have an elevated immunoglobulin G (IgG) response to spirochete, however, Steere (2001) observed that a few patients who receive inadequate antibiotic therapy during the first several weeks of infection develop neurological symptoms but are seronegative. This may explain the equivocal IgG immunoblot test in the initial sample following treatment with oral flucloxacillin. However, most patients who have later manifestations of the illness remain seropositive for years.

Investigating a case of facial diplegia can be challenging. However, a careful history, a thorough physical examination and appropriate laboratory investigations will

Case Report

A 43-year-old man presented to his GP with a right facial palsy of 1 day's duration. He was prescribed a 5-day course of oral prednisolone for a diagnosis of Bell's palsy. Two weeks later he presented to the emergency department with a left facial palsy. He had a 2-month history of back pain, generalized myalgia and paraesthesia. There was no history suggestive of any other neurological deficit. There was no history of fever or weight loss. However, he had travelled to the Scottish Highlands 3 months previously and had noticed an itchy red rash over his buttock (approximately 3 cm in size) 4 weeks after his holidays. The rash was diagnosed as cellulitis and he was prescribed a 5-day course of oral flucloxacillin. The patient, however, did not recall having sustained a tick bite or having a characteristic migrating rash.

Examination revealed a bilateral lower motor neurone facial palsy. Detailed examination of the central and peripheral nervous system was otherwise normal. No vesicles were found in either external auditory canal. Tuning fork tests were normal. The initial investigations undertaken in the emergency unit are listed in *Table 1*.

Repeat serology 8 weeks later was positive for *Borrelia burgdorferi* immunoglobulin G (IgG) antibody by both enzyme-linked immunosorbant assay (ELISA) and immunoblot assay. The initial CSF sample was not subsequently examined for *B. burgdorferi* antibody as the sample had been discarded. A repeat lumbar puncture was not considered appropriate.

This patient's right-sided facial weakness showed spontaneous improvement over 4 weeks from the initial presentation by which time his left-sided weakness had improved.

Based on the history of travel to a tick-infested area, together with the neurological and serological findings, a diagnosis of early Lyme neuroborreliosis was made. Although antibiotic treatment does not hasten resolution of 7th cranial nerve palsy associated with *B. burgdorferi* infection, doxycycline was prescribed to prevent further sequelae.

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Table 1. Initial investigations of a case of facial diplegia

	Tests	Results (reference range)
Biochemical	Full blood counts, urea and electrolytes	Normal
	Liver function tests, lipid profile, bone profile	Normal
	Fasting blood glucose	5.5 mmol/litre (3.5–5.5)
	Erythrocyte sedimentation rate	8 mm at first hour (0–10)
	Serum ACE levels	18 u/litre (18–55)
	CSF cytology	WCC 10/ml (all lymphocytes) No malignant cells, no oligoclonal bands
	CSF biochemistry	Protein 1.5 g/litre (0.15–0.45) Glucose 3.9 mmol/litre (2.8–4.2) Opening pressure 10 cm (7–18)
Autoimmune screen	Rheumatoid arthritis latex screen	Negative
	Double-stranded DNA	Negative
	c ANCA	Negative
	Anti cardiolipin antibody	Negative
	Anti thyroperoxidase and thyroglobulin antibody	Negative
Serological	Syphilis serology	Negative
	Lyme serology (*Borrelia burgdorferi*)	ELISA IgG/M antibody reactive IgG immunoblot equivocal IgM immunoblot negative
Radiological	Chest X-ray	Normal
	MRI brain (non-contrast)	Normal

ACE = angiotensin-converting enzyme; ANCA = antineutrophilic cytoplasmic antibody; ELISA = enzyme-linked immunosorbent assay; Ig = immunoglobulin; MRI = magnetic resonance imaging; WCC = white cell count

significantly narrow down an otherwise long list of possibilities.

Figure 1. Erythema chronicum migrans. Annular erythematous plaque with bright red outer order. Lesions without central clearing (as in this picture) can be confused with cellulitis. Most common sites are thigh, groin and axilla.

This case highlights some useful learning points:

- Facial diplegia is a rare clinical entity which should alert the physician to the possibility of an underlying systemic disorder and requires thorough investigation
- Lyme disease should always be considered as a cause of facial diplegia even in the absence of characteristic rash or other neurological manifestations
- Facial diplegia can be an early presentation of Lyme neuroborreliosis. Patients often have lymphocytic CSF pleocytosis even in the absence of meningeal signs (Stanek and Strle, 2003)
- Lyme disease serology should be interpreted cautiously in patients with prior antibiotic therapy since it can interfere with the antibody response. BJHM

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Table 2. Differential diagnosis of facial diplegia

Infection	Lyme disease
	Tubercular meningitis
	Hansen's disease
	Cryptococcal meningitis with acquired immunodeficiency syndrome
Intracranial tumours	Glioma
	Lymphoma
	Carcinomatous meningitis
Neurological disorders	Guillain–Barré syndrome
	Myasthenia gravis
	Motor neuron disease
	Myopathies
Congenital disorder	Mobius syndrome
Miscellaneous	Diabetes mellitus
	Sarcoidosis
	Trauma
	Pontine haemorrhage
	Systemic lupus erythematosus
	Pregnancy
	Human immunodeficiency virus seroconversion illness
	Hymenoptera sting

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