by Estes et al. in 1967. A further 25 cases have been fully described in the literature since then and case reports have appeared in the English and Japanese literature, although full details of the latter are not available. In 1991 Hardie et al. described 19 cases from the UK, this study being the most detailed to date. Although the syndrome has been described in Europe, Asia and the USA, to the best of our knowledge there have been no cases from Africa.

The case described here displays many of the typical features described previously. Onset of symptoms is typically in the third decade, with males and females equally affected. The most striking neurological feature is stereotyped chorea, with marked involuntary orofacial movements that often interfere with speech and swallowing. The orofacial movements are often a combination of chorea and dystonia as in our patient. The vocal tics and grunting were also seen in 9 of the patients described by Hardie et al. The chorea resembles that seen in HD but involves the legs at an earlier stage while the arm and forehead movements tend to be less severe than in HD. Dementia is variable, but affects two-thirds of patients, although detailed neuropsychological testing may reveal cognitive impairment in a higher percentage of patients than previously thought. Detailed neuropsychological testing shows that the dementia has the characteristics of a fronto-subcortical type.

Motor nerve conduction studies are always normal, but sensory nerve action potentials are reduced in about 50% of reported cases. CT brain scans show varying degrees of abnormality from normal to generalised cerebral atrophy, particularly in the striatum and subcortical regions. A normal brain scan in our patient is in keeping with previous reports. In those cases which have come to necropsy, neuronal loss involving the caudate, putamen and pallidum has been described. The genetic basis of neuro-acanthocytosis remains unclear with autosomal-dominant, recessive and X-linked genetic inheritance proposed. Sporadic cases, such as the patient under discussion, have been reported, but their frequency is probably underestimated.

Awareness of this cause of chorea should prompt fresh blood slides to be examined in any patient with stereotyped or patterned chorea. While most patients present with a movement disorder, some well-documented patients with familial neuro-acanthocytosis did not.

Conclusions

Neuro-acanthocytosis is a rare neurodegenerative disorder which is underdiagnosed worldwide. We report here a case seen at Johannesburg Hospital and suggest that the condition occurs in black Africans and should be considered in patients who present with chorea. Atypical features such as vocal tics, orofacial dyskinesia or areflexia may add to the clinical suspicion.

REFERENCES


Accepted 24 Feb 1997.

Hilton's Rest and Pain, Guy's Hospital personalities and South African rugby connection

H Dubovsky

Guy's Hospital occupies a unique position in medical history. John Hilton (1805 - 1879), as anatomist, physiologist, morbid anatomist and surgeon in his classic Rest and Pain, published in 1863 (reissued in 1950), formulated principles for the diagnostic significance of pain and the value of rest in healing. An array of personalities graced Guy's Medical School in that era. The triumvirate of Richard Bright (1789 - 1858), Thomas Addison (1793 - 1860) and Thomas Hodgkin (1798 - 1866) contemporaneously discovered the diseases that bear their names. Sir Astley Cooper, a leading surgeon of his day (1768 - 1841), performed the first amputation of the hip joint before the era of anaesthesia. John Keats (1795 - 1821) qualified as a surgeon at Guy's but, realising his unsuitability of temperament, became a leading English poet. This change of direction caused him anguish and suffering, mainly because of the rejection of his poetry; tuberculosis led to his death in Rome, where he is buried.

Guy's Medical School also allowed South Africans to enter as rugby players before the 1920s, when they were required to qualify overseas.

The rugby connection

South Africans had no difficulty before the 1920s in entering the prestigious Guy's Hospital Medical School as rugby players when they were required to qualify overseas. There existed at the time a keen competitive rugby league among the medical schools. At a dinner celebrating 250 years of Guy's Hospital in 1976, Sir Hedley Atkins, KBE, Professor of Surgery, commented: 'In 1920 Guy's had a most remarkable rugby football side. It was captained by dear George Doherty, who was alleged to be the only person on the side who spoke English — the rest spoke africans (sic). There was Myburg, and Mostert, and Krige and Albertijn (sic), and Bekker and Van Schalkwijk (sic) ... They all spoke Africans (sic) ... . As a matter of fact, that is not absolutely
true because there was one chap, Graham-Davies, who spoke Welsh. He doesn’t really form an exception because, although he was an international centre-three-quarter for Wales, he only occasionally played for the first team. . . .

When I came here as a student . . . the demands for entry were . . . first that your father was a Guy’s man, the next that you were reasonably competent at rugby and the third, and very much the last, was if you had any academic qualifications.’

Hilton’s Rest and Pain
This work, originally published in 1863 with the title page inscription The Influence of Mechanical and Physiological Rest in the Treatment of Accidents and Surgical Diseases and the Diagnostic Value of Pain, was a series of lectures delivered to the Royal College of Surgeons from 1860 to 1861. The aforementioned H J B Atkins, Chief Surgeon of Guy’s, was co-editor of a 1950 edition. Each lecture is followed by an appendix by the editors, comparing Hilton’s views to current concepts.

John Hilton, FRS, FRCS, started at Guy’s as a demonstrator of anatomy. He was fortunate to have Joseph Towne (1809 - 1879) preserve his fine dissections in wax. Towne started at Guy’s at the age of 17, and had a natural genius for this art, which was essential at a time when anatomical study mainly relied on corpses stolen from graves. Towne insisted on total seclusion when making his accurate colour wax models of dissections and skin conditions. These are on view in the Guy’s Hospital Museum and his technique has remained his secret.

At that time, anatomy and physiology were taught concurrently, influencing Hilton’s observation that the muscles activating a joint and its overlying skin are supplied by branches of the same nerve. Thus, when a joint became inflamed, referred pain to the skin caused the muscles controlling the joint to go into spasm, protecting the joint from further injury. This theory became known as ‘Hilton’s Law’ and earned him a Fellowship of the Royal Society. Rest and Pain was known as the ‘Guy’s man’s Bible’ and emphasised Hilton’s principle that ‘rest is a most important therapeutic agent in the cure of accidents and surgical diseases’. He adds that ‘every pain has its distinct and pregnant signification’ with ‘pain the monitor and rest the cure’. His treatment consists essentially of the immobilisation of affected joints for several months, particularly those that are ‘strumous’ (tuberculous). This was achieved with sandbags, metal strips, leather supports and wooden splints, with adjustable angulation for joints, depending on stage of disease. He administered chloroform, discovered 13 years previously, to relax joint spasm so that optimal angles of joint rest could be determined. Hilton advises on the draining of abscesses to ‘secure coaptation of the internal surfaces to give the surfaces rest so as to permit of their union’. He advises that these be opened at the lowest point to secure this union and not to ‘plunge in a knife’. After an initial skin incision, he uses a blunt dressing forceps to open up the abscess and leave a draining sinus. A feature of Rest and Pain is the accurate line drawings of surgical and morbid anatomy, instruments, fixation appliances and clinical conditions.

Guy’s famous men³
Sir Astley Cooper (1768 - 1841) was responsible for the Anatomy Act of 1832 whereby medical schools could legally acquire bodies for dissection. Accounts stress his attractive personality to contrast with the general image of surgeons of his day who ‘tended to be bluff, hearty extroverts with rude manners and coarse tongues, and walked the wards with their hats on, followed by a cackle of jostling, rather noisy apprentices and pupils’. His elegant and humane operative technique impressed the usually crowded theatre. Operating without the benefit of anaesthesia he successfully ligated the aorta and was the first to amputate the hip joint.

Richard Bright (1789 - 1858), with Addison and Hodgkin, form the famous triumvirate who, while together at Guy’s, discovered the diseases that bear their names. Their pathological specimens are on display at Guy’s Hospital Museum. They were graduates of the Edinburgh school, with its bedside teaching and postmortem correlation of clinical findings. Indeed, it was Bright’s postmortem investigation of albuminuria that led to his discovery of glomerulonephritis in 1827; the involvement of the kidneys in albuminuria had been missed in the past, given that the kidneys were not examined at postmortem. Bright was a fine scientist, and published The Geology, Fauna and Flora of Iceland after participation in an expedition while a student.

Thomas Addison (1793 - 1860) started his career as a dermatologist, and described keloid before his appointment as a physician to Guy’s in 1837. His dermatological knowledge assisted his observation of the skin pigmentation in Addison’s disease of the suprarenals, which he described in 1849. His discovery was not accepted in his lifetime. A sufferer from severe depression, Addison resigned his lectureship at Guy’s, at which he excelled, and committed suicide a few months later by throwing himself from a window.

Thomas Hodgkin (1798 - 1866) started out as a lecturer in morbid anatomy at Guy’s. In 1827, 5 years before Corrigan’s description of aortic regurgitation, he described ‘retroversion of the valves of the aorta . . . which allows of their dropping in towards the ventricles, instead of effectively closing of the vessels’. His application for a physician’s post was refused, probably because he was a Quaker and regarded as a dissenter by the Anglican church. The Quakers did pioneering work in alleviating a wide range of contemporary medical and social problems. Hodgkin’s description in 1837 of the lymphoma that bears his name did not gain acceptance. Sir Samuel Wilks (1824 - 1911), also a Guy’s physician, described the condition 30 years later, but acknowledged Hodgkin’s prior discovery and suggested that the condition should carry his name.

Hodgkin resigned his post at Guy’s but his attempt at private practice was not successful. Fortunately he became full-time personal physician to a retired broker, Moses Montefiore, who devoted himself to the relief of oppressed minorities, both Jews and Christians, in Europe, North Africa and the near East. On one of his visits to Palestine with