Observations on Indian Neurology

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The common neurological diseases of the West are, with some exceptions, frequently seen in Indian practice, but a further catalogue of neurological affliction is associated with the twin factors of infection and malnutrition, and these conditions must feature prominently in any account of neurology in India. Headache, epilepsy, cervical spondylosis, cerebrovascular disease and Parkinson's disease will dominate a neurology clinic anywhere in the world, but the Indian neurologist will also see many other diseases regarded as exotic in the UK and as commonplace in India. The first generation of immigrants to the UK from the Indian sub-continent, and to a lesser extent later generations, continue to manifest some of these conditions and, in urban areas of the UK with a high immigrant population, the clinician must therefore consider these unusual diagnoses. These points are illustrated by a recent study from Leicester[1] in which it was noted that 15 of 17 cases of tuberculous meningitis (TBM) were in Asian patients and that late diagnosis was sometimes a problem. This article highlights some of the neurological diseases which lie outside the routine experience of the English trained doctor, but are familiar to his Indian colleague.

Tuberculosis of the Nervous System

Tuberculous Meningitis (TBM)

The incidence of TBM in the UK is of the order 1.5–2.0 cases per million per year[2,3]. Comparable figures for India are not available but a busy neurology unit will see well over 100 cases each year and many more patients will be treated by general physicians and paediatricians. There is no doubt that TBM is over-diagnosed in India; this is inevitable where isolation rates are low[4] and when treatment is urgent. However, late diagnosis can be a problem in the UK, where clinicians may not think of TBM, particularly when the presentation is atypical. The overall clinical picture, laboratory findings and prognosis are similar in India and the UK, but some differences are seen. The Indian patients are more likely to be children, more likely to have miliary or pulmonary disease, and a history of direct tuberculosis exposure is frequently obtained. The less usual presentations of TBM are seen in abundance in India, for instance subacute blindness due to chiasmatic and optic nerve arachnoiditis, and acute hemiplegia in which meningeal disease may have occluded major cerebral vessels. The treatment protocols in India have concentrated on the less expensive antibiotics, often without rifampicin, and it is therefore interesting that morbidity and mortality statistics do not differ significantly between the UK and India[3,5,6] and are very similar to those obtained in the early days of anti-tuberculous chemotherapy[7]. It is surprising that we are still unclear about the indications for steroid treatment and intrathecal therapy, but prompt initiation of standard anti-tuberculous treatment as soon as the diagnosis is suspected is the main guarantee of a successful outcome.

Intracranial Tuberculoma

In the UK, intracranial tuberculoma is usually seen in recent immigrants, although rare cases appear in the indigenous population[8]. In India, this condition is common and studies of incidence in neurosurgical units gives a range of 4.1–30.5 per cent of all intracranial tumours[9,10]. Now that CAT scanning facilities are becoming widespread in India, more cases are identified at an early stage, multiple lesions are frequently found and the response to a combined medical and surgical approach, or to medical treatment alone, can be monitored precisely[11]. The clinical features of intracranial tuberculoma are similar in both countries, lesions may be found in the background of a known tuberculous infection or the presentation may be that of any cerebral space occupying lesion, with a progressive neurological deficit in the presence or absence of symptoms of raised intracranial pressure.

Adhesive Spinal Arachnoiditis

Adhesive spinal arachnoiditis of presumed tuberculous origin accounted for nearly 10 per cent of paraplegia in a large representative Indian study[12]; a British neurologist will occasionally see this entity. The patient will characteristically be Indian, but of the three cases seen in the UK by one of the present authors, two have been young English males. The condition may arise by spread from a tuberculous basal meningitis or less commonly from spinal tuberculous caries. A subacute progressive myeloradiculopathy, the primary spinal type, is also seen and here a wide differential diagnosis must be exercised[13]. Myelography by lumbar or cisternal route and spinal fluid analysis are mandatory, and while treatment

of the likely underlying active tuberculosis is important, medical and surgical attempts to reduce the deficit due to the arachnoiditis have not met with great success. Recent work with hyaluronidase has suggested a further treatment option in both spinal[14] and cranial[15] arachnoiditis, although further studies are necessary to evaluate the therapy.

Neurological Complications of Leprosy

Mycobacterium leprae has a peculiar predilection for the peripheral nervous system, but in India as elsewhere these problems are usually dealt with by the leprologist rather than the neurologist. Nevertheless, a case can be made for the more active involvement of neurologists in the study and treatment of this disease. The Ridley-Jopling classification[16] subdivides leprosy into the polar types of tuberculoid (high resistance) and lepromatous (low resistance), their borderline variants and the intermediate forms. The peripheral nervous system is involved in all types of the disease and there is also a controversial pure neuritic variant, in which careful enquiry and examination fail to provide evidence of cutaneous lesions. The facial, ulnar, median, radial and common peroneal nerves are frequently affected in leprosy and the Indian patient with a Bell's palsy or ulnar neuropathy must be meticulously examined for the presence of cutaneous lesions or nerve thickening. Conversely, in Indian practice, the temptation to diagnose leprosy without adequate clinical grounds or biopsy must be resisted, as cases of amyloidosis, porphyria, congenital insensitivity to pain and especially hereditary sensorimotor neuropathy may all appear neurologically similar to leprosy. The tragedy is that such patients may be sent to a leprosy establishment and thereby become exposed to the disease. The treatment of leprosy with dapsone and the newer antileprotics will arrest the infection, but regrettably continuing fibrosis within the nerve may increase the deficit despite otherwise successful treatment, and repeated trauma to anaesthetised extremities may result in dreadful deformity.

Cysticercosis

Cerebral cysticercosis is endemic in the non-Muslim populations of the developing countries and India is no exception[17]. The presence of the cysts of *Taenia solium* in the brain usually results in one of three symptom groups. The patient may exhibit features of raised intracranial pressure, a mental disturbance or convulsions, or a combination of these. Occasionally the disease is identified on radiological examination for other complaints, and rarely cysts may be found in the brain of autopsied patients who have died from other conditions and shown no indication of cysticercosis in life. If cysts are present in the muscle of human hosts a striking muscular hypertrophy may be seen, and the calcified lesions are readily observed radiologically. The treatment of cysticercosis with drugs or surgery is often unsatisfactory and control of this disease will only be possible through effective public health education.

Virus Infections of the Central Nervous System

Poliomyelitis remains a relatively common disease in India. Vaccination programmes are widespread, but in this population more booster doses of vaccine than those given in the West appear to be necessary to achieve adequate immunity[18]. Cases occur sporadically in the UK but, with the present general trends against vaccination and towards more widespread foreign travel, it seems possible that poliomyelitis may re-emerge and it is wise to consider this infection in the differential diagnosis of cases of post-infective polyneuropathy.

Acute haemorrhagic conjunctivitis due to infection with enterovirus 70 has associated neurological features that are in many ways similar to those of poliomyelitis. Widespread epidemics were seen in India in 1971 and 1981, male adults were predominantly affected, and the clinical features were a rapid paralysis of the limbs and the cranial nerve innervated musculature. A detailed study[19] has suggested that the major neurological lesion is at the level of the anterior horn cell, and a permanent poliomyelitis-like disability may follow the acute illness.

Japanese encephalitis caused by a Group B arbovirus occurs in epidemics but is also endemic in parts of India, particularly the south. This mosquito-borne infection finds a reservoir in pigs, cattle and some species of bird. A prodromal phase is followed by an acute encephalitis from which only one-third of affected individuals recover, a further third remain neurologically damaged and one-third die[20]. A vaccine is available and, as with acute haemorrhagic conjunctivitis, control of these devastating conditions is theoretically possible. Recently, a tick-borne *epidemic encephalitis* occurring in the forests of the State of Karnataka has attracted attention. Having bitten infected monkeys, the ticks fall to the forest floor to await the next meal from an unsuspecting human host, in whom haemorrhagic complications often prove fatal.

Other Neurological Infections

In a single year there were 227 cases of *tetanus* seen at the J. J. Group of Hospitals, Bombay[21] and 70 of these patients died of the disease. Improved supportive care may now be reducing the mortality but tetanus remains an important but preventable neurological infection.

The scourge of *rabies* continues in India; few have access to diploid cell anti-rabies vaccine. Imported cases may not become unwell until after entry to the UK[22].

Neurosyphilis, while declining in incidence, may still be seen, especially in the acute or subacute meningovascular forms, but many Indian neurological departments may not have one case in a year. There is an impression that *subacute sclerosing panencephalitis* is more common in India than in the UK, and it is possible that a widespread measles vaccination programme may reduce the prevalence of this 'slow virus' disorder.

Non-infective Spinal Cord Disorders

In South India the most common form of myelopathy is a subacute spastic paraplegia with peripheral sensory

symptoms, dorsal column involvement and, frequently, bladder disturbance[23]. Identification of known causes of paraplegia will allow categorisation of a few cases, but a large enigmatic group of patients remain. There are similarities to the tropical paraplegia seen elsewhere, notably in Jamaica and Africa, but in all cases intensive study has failed to reveal a nutritional, toxic or infective cause for this entity.

Lathyrism

Lathyrism, an acute or subacute corticospinal tract degeneration, is common in Central India, and may be found in 4–6 per cent of landless labourers in endemic areas. This disease follows the consumption of the seeds of *Lathyrus sativus,* and was recognised by the ancients, as illustrated by the 'immortal verse' of lathyrism, thus 'the black pea with its yellow flower, from eating it, comes trouble in the legs, flopping top knot and swaying hips: behold the ill effects of eating matra'[24]. This easily grown crop is used when there is a shortage of conventional food but detoxification of the pulse is possible and health education could banish this disorder to the realms of medical history.

Endemic Fluorosis

The high concentration of fluoride in the drinking water of many parts of India may result in endemic fluorosis, which is particularly likely to occur when the water source is further concentrated by evaporation. Mottling of the teeth is an early sign, but a generalised osteosclerosis may become neurologically manifest when the spinal cord and spinal roots are compressed by vertebral disease[25]. Once again, the major prospect for prevention of this condition lies in appropriate public health activity, in this case the provision of unaffected water supplies, which poses insurmountable engineering and financial difficulties in a country as vast and populous as India.

Atlanto-axial Dislocation

In Western practice, disease of the atlanto-axial joint is likely to be associated with rheumatoid arthritis, but in an Indian population a congenital form of atlanto-axial dislocation is relatively common. This entity may present with cervical pain and stiffness, transient attacks of neurological disturbance or a progressive spinal cord syndrome. Clinical suspicion can usually be confirmed by plain radiology of the cervical spine with attention to the distance between the odontoid and the anterior arch of the atlas, and to the dimensions of the cervical canal, but myelography may sometimes be necessary when the diagnosis is not clear. Surgical reduction of the dislocation and lateral fusion are successful, especially in early cases[26].

Other Neurological Diseases common in India

Painful ophthalmoplegia, also referred to as the superior orbital fissure syndrome or the Tolosa-Hunt syndrome, is disproportionately common in India and a para-infective aetiology has been postulated where an association with past tuberculous or filarial infection has been found [27]. This may be fortuitous and, as in the much rarer UK cases, a dramatic response of the facial pain, ophthalmoparesis and raised erythrocyte sedimentation rate is seen following corticosteroid treatment.

Classical *motor neurone disease* occurs in India with a frequency similar to that in the UK, but an unusual juvenile variant, with onset before the end of the third decade, is seen in India, particularly the south[28]. Bulbar and asymmetrical limb involvement is characteristic and a more benign prognosis can be anticipated. The involvement of the pyramidal tracts and the absence of a family history differentiate this group from cases of spinal muscular atrophy and no association with the polio virus has been found. A further unusual form of motor neurone disease, a monomelic variant, has also been observed, and a number of Indian cases have recently been studied in detail[29].

Cerebrovascular disease afflicts the Indian to the same degree as the Westerner, but there is an impression that strokes in younger patients may be more prevalent in India. An obliterative aortic arch syndrome appears to be quite common and it is possible that a greater proportion of strokes are due to an underlying arteritis, rather than to the familiar embolisation from large vessel arteriosclerosis[30]. Conversely, it was thought that intracranial aneurysm was less common in India than elsewhere, but this observation most likely reflects only the difficulty in collecting statistical data in India. Large scale epidemiological studies are under way, both in cerebrovascular disease and in wider aspects of neurology, and the results will be of great interest.

A fascinating form of reflex epilepsy has been described in which partial or generalised seizures are induced by bathing with hot water. In a large series of patients with epilepsy[31], 108 (9 per cent) were found to have *hot water epilepsy* in which attacks were precipitated by pouring mugfuls of hot water on to the head and body, a popular method of bathing in India. This remarkably high frequency contrasts with the very rare Western case reports of hot water immersion epilepsy[32]. Treatment may simply be to use tepid water for bathing, although anticonvulsants have been used in some cases.

A mention should be made of the high incidence of some of the genetically-mediated neurological disorders in India. *Spino-cerebellar degenerations* are prevalent in some areas, and *mental handicap,* caused by obstetric and genetic factors, is a major health problem. *Wilson's disease* shows remarkable clustering. The personal series of one neurologist in India, collected largely from the local population, stands at 52 cases[33]. It is possible that in rural areas consanguinity is an important factor in the determination of these conditions.

Conclusion

Neurological disease varies enormously worldwide and this brief review has detailed some of the conditions which are commonly seen in India and the Indian, but rarely

appear in a British consulting room or ward. The reasons for the high incidence of infective and nutritional disease in India are obvious, but there is no known reason for the superior orbital fissure syndrome or hot water epilepsy being so common in the practice of an Indian neurologist and so rare in the UK. The study of comparative neurology affords fascinating glimpses into the aetiology of these conditions, and the neurologist in training as well as the established clinician should take every opportunity of examining his subject in another environment.

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