Review

Disappearing neurological diseases in Sri Lanka

J B Peiris[1]

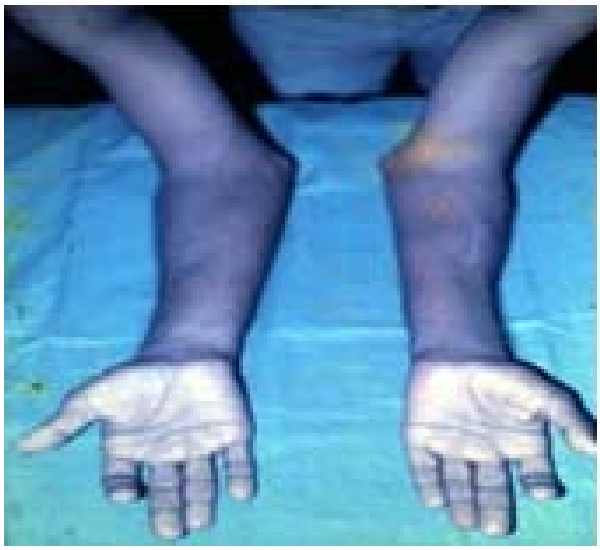
Sri Lanka Journal of Neurology, 2014, **3**, 16-19

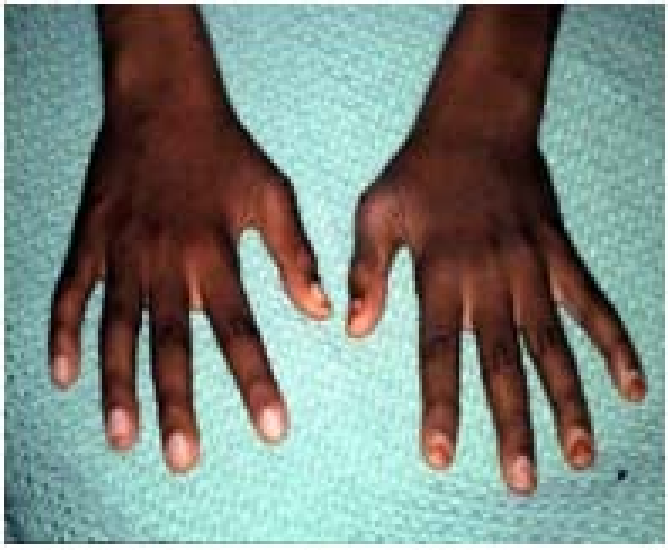
Disappearance of a disease is a rarity. Arguably, the only disease to have been erased from the face of the earth is smallpox. Poliomyelitis is hopefully the next in line, but in spite of a concerted effort worldwide, civil strife and political interference, are preventing the eradication of poliomyelitis in a few countries. Many neurological diseases are chronic with little hope of recovery, leave alone worldwide eradication. In this context even a reduction of incidence, in a localized region is significant and deserves a closer look.

The diseases to be described are seen mostly in Asian countries and that too in the last 50 years. Some have been documented only in Sri Lanka.

Distal spinal muscular atrophy (DSMA)

My first contact with DSMA was in 1972. The first patient that drew my attention was a 21 year male with insidious onset, non-familial, bilateral, asymmetrical, wasting of his hands and forearms. There was no lower limb, bulbar, sensory or sphincter involvement. There were no long tract signs. Hitherto, similar patients had not been seen in the country. During my 2 years training in London, Edinburgh and Glasgow, I did not see a single patient with similar manifestations. For a diagnosis, rare conditions like distal myopathy, a pure motor neuropathy and a cervical myelopathy were considered, though they were clinically only remote possibilities. In the next 3 years, 33 patients were seen with similar presentations. There was a remote resemblance to Madras Motor neuron disease. This made me consider it a motor neuron disease in the young and presented it as such for the silver jubilee celebrations of the Institute of Neurology, Madras[1]. As the years passed and more patients were seen with a longer follow up, its resemblance to the Japanese Hirayama disease-atrophy of the distal upper extremity became apparent, though there were a few distinct features[2]. Over 100 more patients were seen over the next 10 years, when our series had more patients than that seen in Japan[3]. Affection of the upper arm and shoulder was rare. Most patients had a spontaneous arrest in 5-7 years. New patients with similar symptoms are now rare and the reason is obscure. Over the last 10 years, the numbers have dropped significantly amounting to less than 10 annually.





[1] *Senior Consultant Neurologist, Colombo, Sri Lanka.*

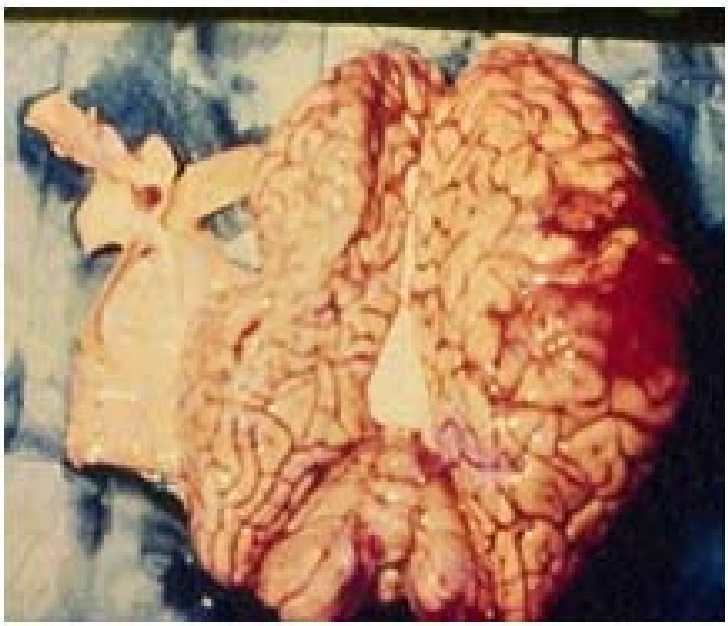
No definite cause for this entity was found and absorption of a pesticide through the skin of the hands was postulated, and the spontaneous arrest may have been due to discontinuation of agricultural activity though in some an exposure to pesticides were not present. This was during an era when mechanized agriculture and routine use of gloves were not common.

Retinopathy in megaloblastic anaemia

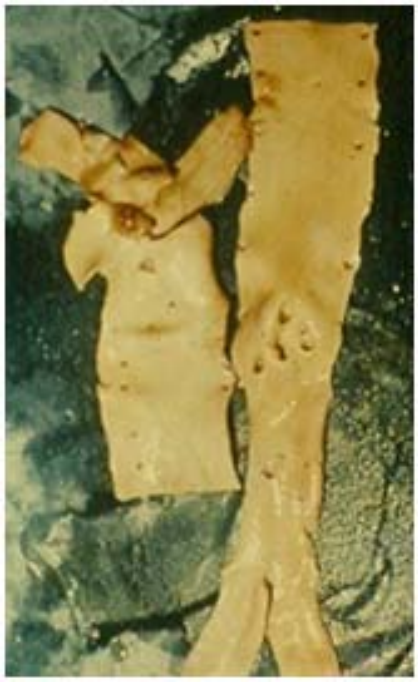
In the same location, the University Unit of Peradeniya (where I was registrar), around the same time as my encounter with the first patient with DSMA, there were many megaloblastic anaemia patients from the tea plantations developing a retinopathy. The retinal haemorrhages and exudates disappeared in 7-10 days of treatment – with vitamin B12 initially and with folic acid, if there was no response. The anaemia was accompanied by a depressed leucopoiesis with few megakaryocytes and diminished platelet formation in marrow. A comparable group of 20 iron deficiency anaemia patients had no thrombocytopaenia and no retinopathy. There has been a paucity of publications on retinopathy in megaloblastic anaemia, but none from Sri Lanka in the last 45 years. The improved nutrition of the tea plantation workers may have been responsible for the virtual disappearance of megaloblastic anaemia and accompanying retinopathy[4].

Transient emboligenic aorto-arteritis (T E A)

Strokes in the young without the accepted risk factors of hypertension, hyperglycaemia, hyperlipidaemia, obesity, smoking and alcohol abuse, appear to be a problem common in Asian countries. The advent of echo cardiography, duplex carotids and Holter monitoring has unearthed a few young stroke patients with a treatable cause but there is still a significant number without a cause. By doing immaculate autopsy studies on 10 young stroke patients who died, the neuropathologist in General Hospital, Colombo was able to identify unusual hitherto undocumented lesions in the aorta and cervical vessels which embolised to the cerebral circulation producing infarcts. There were transient inflammatory lesions in the media with an overlying thrombus which embolised. The lesions in the media were transient leaving a scar which could easily be missed, and are unlikely to be detected by sonography or arteriography. Hence, even in this day and age, with all the modern imaging techniques available, the condition is likely to be missed unless focused and detailed autopsies are carried out, with special staining of suspect healed areas[5].

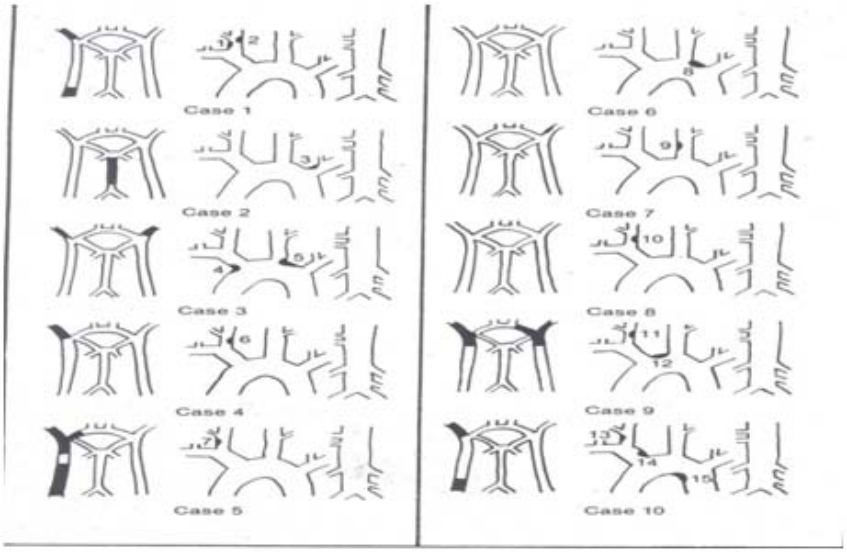


Brain of patient with haemorrhagic infarct of R. hemisphere and scar of old infarct in L.



Arch of aorta with fleshy thrombus and scar of old.

Circle of Willis Aortic arch Desc aorta



- All 10 autopsies showed an intra-cranial thrombotic lesion
- All showed 1 or more lesions in aorta or thoraco-cervical vessels

Rest of aorta showed no atheroma

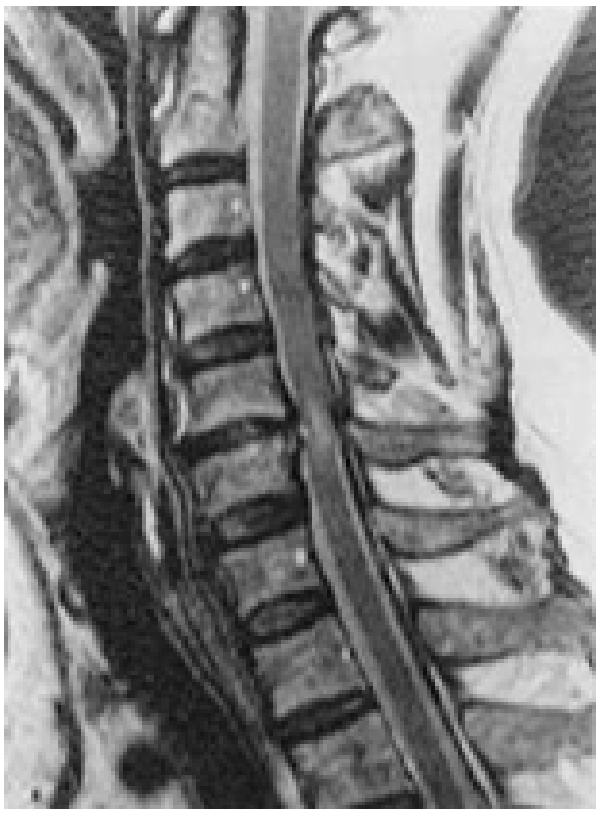
Juvenile cervical spondylosis

Cervical spondylosis is essentially a disease of ageing and is uncommon under the age of 40 years. In a short space of 9 months, five patients under the age of 40 years were seen with recent onset, insidious, spastic paraparesis of 1-6 months with soft signs in upper and lower limbs – impaired or inverted supinator jerk, slightly exaggerated triceps and knee jerks. Some gave a history mild indirect cervical trauma.

Clinical diagnosis was a non-compressive cervical myelopathy and the MRI showed a mild C3/C4 or C4/C5, disc protrusion. The neurosurgeon was persuaded to do a discectomy even without 'adequate indication' before dooming the patient for a lifetime of gloom. At surgery, one or more soft cervical discherniations were found and patients underwent cervical discectomy with or without bone grafts. Patients made a good recovery and returned to normal life[6].

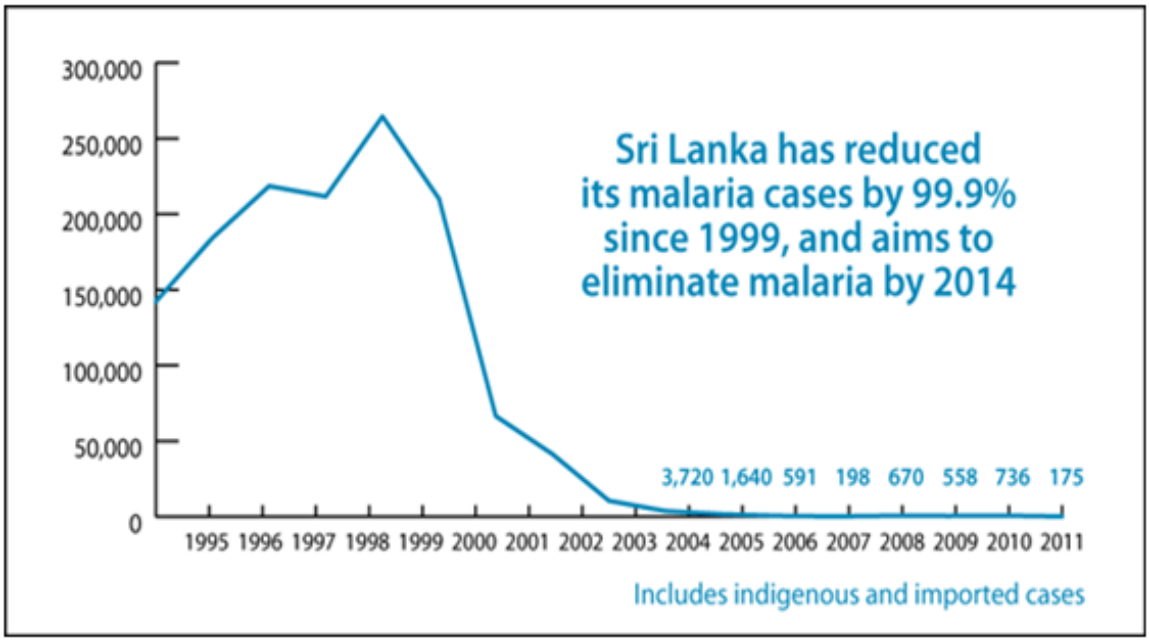
The neurosurgeon who operated on our patients migrated to Malaysia and continued the study in Malaysia and described 26 patients with disabling symptoms of less than 6 months, 58% presenting with a spastic paraparesis. Few had bladder symptoms and motor symptoms in upper limbs and may have presented later than the Srilankan counterparts[7].

No patients have been described in Sri Lanka over the last 30 years – it is possible that some are being missed, due to non-consideration of it in the diagnosis, even with greater availability of MRI scanning.



Cerebellar malaria

A 40-year-old female was transferred from a malarial endemic area for exclusion of a posterior fossa tumour by CT scan because of a disabling truncal ataxia. Examination revealed midline cerebellar disease without raised ICP, brain stem, CP angle or long tract signs.



There was no clinical evidence to suggest a space occupying lesion. A focused detail history revealed that there were others in the same locality with almost identical manifestations. Malaria was rampant in the area at that time and many had taken multiple doses of anti-malarial drugs and had multiple episodes of malaria including falciparum malaria. We were uncertain as to whether it was a hitherto undocumented effect of falciparum malaria or was it an unrecognized form of toxicity to multiple doses of anti-malarials – mostly chloroquine[8]. Fortunately we were able to establish a link with Oxford university and we concluded that it was due to immune activation causing cerebellar dysfunction following plasmodium falciparum malaria[9]. Malaria has been at a single digit level since 2004, and is now completely eradicated, and with it cerebellar malaria.

References

1. Peiris JB, Seneviratne KN, Wickremasinghe HR. Motor Neuron disease in the young *Proc. Inst of Neurology, Madras* Silver Jubilee edition, 1976; **6**: 160.
2. Hirayama K, Tokokura Y, Tsubaki T. Juvenile muscular atrophy of unilateral upper extremity. *Psychiatry and Neurology* (Japan), 1959; **61**: 2190-7.
3. Peiris JB, Seneviratne KN, Wickremasinghe HR, Gunatilake SB, Gamage R. Non familial Juvenile Distal spinal muscular atrophy of upper extremity. *J of Neurology, Neurosurgery and Psychiatry* 1989; **52**: 314-19.
4. MacanMarkar MA, Peiris JB, De Silva GU, Prematileka, N. Transactions of the Royal Society of Tropical Medicine and Hygiene 1969; **63**: 398-406.
5. Wickremasinghe HR, Peiris JB, Thenabadu PN, Sheriffdeen AH. Transient Emboligenic Aorto-arteritis – a noteworthy new entity in the young stroke patient. *Archives of Neurology* 1978; **35**: 416-22.
6. Peiris JB, Selladurai BM. Compressive cervical myelopathy in the young due to multiple soft disc protrusions. *Ceylon Medical Journal* 1981; **26**: 59-63.
7. Selladurai BM. Cervical myelopathy due to nuclear herniations in young adults: clinical and radiological profile: results of Microdiscectomy, without interbody fusion. *Journal of Neurology, Neurosurgery and Psychiatry* 1992; **55**: 604-08.
8. De Silva HJ, Peiris JB, Gamage R, Herath HKN, Abeysekara DT. Tropical Doctor. A self-limiting midline cerebellar syndrome – is Falciparum malaria the cause? 1988; **18**: 5-6.
9. De Silva HJ, Hoang P, Dalton M, De Silva NR, Jewell DP, Peiris JB. Immune activation during cerebellar dysfunction following plasmodium falciparum malaria. *Transactions of Royal Society of Tropical Medicine and Health* 1992; **86**: 129-31.