Letters

ticularly with gram negative organisms is a well recognised complication of strongyloides hyperinfection. In the review by Igra-Siegmán *et al.*⁸ 44% of cases developed secondary bacterial infection. Eleven per cent had gram negative meningitis mainly from *E coli* infection, and all died. This secondary infection appeared to be the result of strongyloides hyperinfection rather than a coincidence of two infections precipitated by immunosuppression, as there was no difference in the incidence of secondary infection between those who were immunocompromised and those who were not. Enteric bacteria are thought to gain access to the subarachnoid space from the blood streams following ulceration of the bowel or they may actually be carried into the meninges by larvae that enter the blood.

Hyperinfection of immunocompromised patients with strongyloides has an 86% mortality rate.⁸ The development of secondary bacterial meningitis is almost universally fatal with only two documented survivors.⁷ Since the hyperinfection syndrome may be precipitated by immunosuppression, it is imperative that strongyloidiasis be sought by stool examination in anyone with a history of previous residence or travel in an endemic area.²⁴ Initial stool examination may be negative, as in our patient, and should be repeated periodically during immunosuppressive therapy.¹⁵ Disseminated strongyloidiasis infection should also be considered in all patients presenting with *E coli* meningitis who have been resident or travelled at any time in an endemic area. In our patient, a fatal sequel to steroid therapy might have been prevented by regular stool examination. Increased awareness of this hyperinfection syndrome is necessary if future deaths are to be prevented.²⁵

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Extensor tone disinhibition from an infarction within the midline anterior cerebellar lobe

Sir: The regional physiology of the anterior cerebellar lobe (ACL) is unclear in man; small cortical lesions are rare, and precise clinical-anatomical correlations in lower primates are not available for comparison.¹ In classic experiments on whole cats, unilateral vermal cortical lesions produce marked ipsilateral limb, back, and neck extension in association with contralateral limb flexion. Bilateral vermal cortical lesions produce marked extension of all four extremities, opisthotonus, and a positive supporting reaction. Midline ACL neurons have ipsilateral cerebellar cortico-vestibular inhibitory efferents to the lateral vestibular nucleus of Deiters, and appear to inhibit ipsilateral muscle tone. The net effects of midline ACL injury in cats sparing deep nuclei are the loss of inhibitory tone upon the ipsilateral vestibulospinal tract neurons within the lateral vestibular nucleus of Deiters, and subsequent disinhibition of extensor muscle tone.^{6,8} Although similar cerebellar cortico-vestibular projections are present in lower primates, precise clinical-anatomical correlations have not been made with small cortical lesions.^{7,9,10} To our knowledge the following case represents the first reported patient with marked extensor rigidity from an infarction within the vermal and paravermal cortex of the ACL, and suggests that vermal and paravermal cortical neurons also inhibit extensor tone in humans.

A 47 year old woman, in good health, suddenly developed ataxia and headache while doing household chores. Initial examination revealed a pulse of 80 beats per minute, blood pressure 150/80 mm Hg, normal fundi, absence of vascular bruits,

and no murmur. The remainder of the general examination was normal. Neurological examination revealed a normal mental status, intact cranial nerves (including normal pupillary size and reflexes), intact sensory systems, and plantar flexion responses. Cerebellar testing revealed mild dysmetria and dyssynergia in the left upper extremity. There was marked spontaneous extensor posturing of the neck and trunk, sometimes appearing opisthotonic. Supine, all four extremities exhibited increased extensor tone and hyperreflexia without clonus. The extensor activity was enhanced in the erect posture with rigid extension of the legs, and extension and pronation of the feet. Walking was difficult owing to the marked extensor tone that appeared as an exaggerated supporting reaction. Heel walking could not be performed.

Initial computed tomography was normal, but 5 days after admission a left vermal and paravermal infarction was noted in the anterior cerebellar lobe. The infarction was approximately 2.0 cm in depth, did not extend into the deep cerebellar nuclei, and

was probably from occlusion of a median branch of the superior cerebellar artery (fig). After one week the symptoms resolved and the patient was discharged. Magnetic resonance imaging was not available before the patient was lost to follow up.

Symptoms from cerebellar dysfunction are generally associated with large subdivisions, probably because of the increased frequency with which these areas are affected. Deficits of voluntary limb movements are related to hemispheric lesions, postural abnormalities such as truncal ataxia to midline lesions, and eye signs such as nystagmus to the flocculonodular lobe.³ Smaller subdivisions, such as the midline ACL cortical neurons, appear important in that they inhibit muscle tone experimentally. Subprimate experiments with cats demonstrated enhanced extensor tone of the extremities, contraction of back muscles including opisthotonus, and positive supporting reactions. The latter followed destruction of vermal neurons and their cortico-vestibular inhibitory efferent projections to the ipsilateral vestibular

nuclei. Bilateral vermal cortical destruction appears necessary to produce hyperextension of all extremities in subprimates.^{6,8}

In primates the function of midline ACL neurons is less clear. Studies in lower primates have demonstrated ipsilateral cerebellar cortico-vestibular, ipsilateral cortico-fastigial, and bilateral fastigio-vestibular efferent projections from the midline ACL.^{7,9,11}

Clinical symptomatology was not noted in the above studies, however, since the lesions were very small, approximately 2.0 mm by design. Hyperextension of the limbs was observed in one study with monkeys, but was produced by a bilateral vermal lesion.¹⁰ Since lower primate fastigial nuclei have bilateral, symmetrical efferent projections to both lateral and inferior vestibular nuclei, it is possible that the cortico-fastigial and fastigio-vestibular connections act in series to influence extensor tone. With the loss of unilateral ACL cortical inhibition upon ipsilateral fastigial neurons and subsequent bilateral efferent input upon the lateral vestibular nuclei, extensor tone may be increased bilaterally. Thus, a unilateral vermal and paravermal infarction may produce bilateral increased extensor tone. Conversely, ischaemia surrounding the observed infarct may have affected superior ACL neurons bilaterally. The net result would again be bilateral disinhibition of extensor tone from bilateral loss of cortico-vestibular inhibitory efferents with or without contribution from a cortico-fastigial and fastigio-vestibular connection. Clinical signs, in either case, would include hyperextension of all extremities and neck, opisthotonus, and a positive supporting reaction.

Extensor tone disinhibition probably does not occur in most ACL lesions since midline cortico-vestibular and cortico-fastigial systems are not selectively destroyed. Additional involvement of the deep midline nuclei would also eliminate efferent fastigio-vestibular output and potentially result in decreased truncal tone and ataxia. Careful review of the original literature regarding alcoholic cerebellar degeneration indicates neuronal degeneration and gliosis in the anterior superior vermis, as well as degenerative changes in fastigial, globose, emboliform, and occasionally dentate nuclei.¹² One review did not discuss the deep nuclei in detail and unfortunately reinforces the concept that ACL injury will only result in broadly-based truncal ataxia.¹³ Further unbiased observations will be necessary to characterise the function of ACL cortical

Fig Enhanced CT scan viewing posterior fossa in sequential 5.0 mm levels through the superior cerebellum (A-D). Note the left vermal and paravermal infarction.

neuron subpopulations and their influence upon muscle tone in humans.

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Transient ischaemic attacks associated with thrombocytosis in active rheumatoid arthritis

Sir: Cerebrovascular involvement associated with rheumatoid arthritis is rare. Most of the vascular complications have been related to vasculitis,^{1,2} hyperviscosity syndrome³ and cervical rheumatoid arthritis.⁴ However, thrombocytosis has rarely been reported with cerebral events in patients with rheumatoid arthritis.^{5,6} We describe a patient who suffered from rheumatoid arthritis, thrombocytosis and transient ischaemic attacks.

A 54 year old man with seropositive rheumatoid arthritis since the age of 30 years was admitted because of recent acute exacerbation of rheumatoid arthritis and two episodes of paraesthesia of the left cheek and arm and dysarthria followed by complete improvement in minutes. Previous therapy had included prednisone, gold salts and indomethacin. General examination showed findings typical of active rheumatoid arthritis. Neurological examination was normal. ESR was 110/140 mm (Westergren) Hb 9 g/dl, platelets 620,000/mm³ and latex fixation test positive in a dilution of 1/640. CT of the brain, EEG, chest radiograph and ECG were normal. The cervical spine was not markedly abnormal. Both supraaortic and cerebral IV digital subtractions angiography were normal. Aspirin, dipyridamol, indomethacin, azathioprine and rehabilitation were started. Gradually the patient's condition improved, the platelet count decreased to 300,000/mm³ and no more neurological disturbances were observed.

Thrombocytosis is usually asymptomatic; however, it may induce thrombosis by means of platelet aggregates.⁷ Thrombocytosis may occur in patients with active rheumatoid arthritis⁸ but thromboembolic phenomena have rarely been reported.^{5,6} We believe that our patient's symptoms were related to the elevated number of platelets, and can exclude on clinicoradiological grounds both vasculitis and vertebrobasilar insufficiency. The reduction in platelets count we ascribe to administration of azathioprine.

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Notice

8th World Congress of Psychiatry. This will be held in Athens, Greece, 13-19 October, 1989. Information may be obtained from the Congress Secretariat, 4 Eginitou Street, GR-115 28 Athens, Greece.