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Are alpha-1-antichymotrypsin and inter-alpha-trypsin inhibitor peripheral markers of Alzheimer's disease?

The definite diagnosis of Alzheimer's disease (AD) requires both clinical criteria of probable AD and neuropathological evidence of AD lesions.[1] At present there is no laboratory test for a premortem diagnosis. Recently, genetic and histochemical studies identified protease inhibitors as components that might be implicated in the formation of the amyloid substance in AD brains. First, Abraham *et al*[2] suggested a potential role of alpha-1-antichymotrypsin (ACT) in the pathogenesis of the lesions, moreover Matsubara *et al*[3] found an increased serum concentration of ACT in AD. Second, several authors[4–6] showed that one transcript of A4 amyloid precursor contained an additional sequence similar to the active site of inter-alpha-trypsin inhibitor (ITI). The purpose of our study was to test the diagnostic value of ACT and ITI in serum and CSF from AD patients.

Sera and CSF were collected from eight men and 16 women with probable AD,[1] mean (SD) age 66 (9·8) years, and from a control group of 19 men and six women aged 64 (8·3) years. Controls were volunteers free of any neurological disease, with a MMS score higher than 28, who had had a myelo or radiculography for proven disk herniation. CSF was not collected especially for this study. The procedure was approved by the ethical committee of Lille. ACT and ITI contents were measured by electroimmunodiffusion methods. Semi-quantitative determination was used for ITI in CSF because of its low concentration. Statistical assessment used non parametric tests (Mann and Whitney's U test and Spearman's rank correlation test).

In the control subjects there were 1) no difference in serum or CSF ACT and ITI contents between males and females, 2) no correlation between age and both serum ITI and CSF ACT contents, 3) a positive correlation between serum ACT contents and age ($p < 0.02$).

Between AD patients and controls, there were no difference in serum or CSF ACT and ITI contents, and no difference of the ACT CSF/serum ratio (table).

In AD patients there was no correlation between the severity of dementia on MMS and Blessed scores and serum or CSF ACT contents, and a negative correlation between MMS and Blessed B scores and serum ITI contents ($p < 0.05$).

Our results show that ACT and ITI are not useful markers of AD in serum and CSF. They don't confirm those of Matsubara *et al*.[3] The ACT CSF/serum ratio was not significantly modified in AD patients, which is consistent with the hypothesis that the blood-brain barrier is not strongly affected in this disease. The correlation between serum ITI contents and the severity of the dementia could be explained by non specific metabolic disturbances.

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Table Serum Alpha-1-antichymotrypsin (ACT) and Inter-alpha-trypsin inhibitor (ITI) contents, CSF ACT contents and ACT serum/CSF ratio in controls group and Alzheimer's disease (AD) patients.

		Controls group	*AD Patients*
ACT mean (SD)	Serum	0·67 (0·27) g/l	0·63 (0·22) g/l
	CSF	6·97 (1·45) mg/l	7·34 (0·66) mg/l
	Serum/CSF	11·46 (4·5)	12·14 (4·53)
ITI mean (SD)	Serum	0·71 (0·19) g/l	0·72 (0·29) g/l

Postradiation motor neuron syndrome of the upper cervical region—a manifestation of the combined effect of cranial irradiation and intrathecal chemotherapy?

CNS prophylaxis is now an integral part of the treatment of acute leukaemia. We wish to report an unusual case of neurogenic amyotrophy apparently resulting from damage to the anterior horn cells of the upper cervical cord and lower brainstem during cranial irradiation.

The patient presented at the age of 13 in January 1977 with T-cell acute lymphoblastic leukaemia and was treated according to the United Kingdom Acute Lymphoblastic Leukaemia Trial 4 (UKALL 4) (intensive) schedule. This comprised induction with cyclophosphamide, cytosine arabinoside (ara-C), vincristine, prednisolone and intrathecal ara-C; consolidation with the same, together with adriamycin, asparaginase, 6-mercaptopurine, intrathecal methotrexate and cranial irradiation; and maintenance with vincristine, methotrexate, ara-C, 6-mercaptopurine and prednisolone. The total dose of irradiation was 2400 cGy (rads) and the field extended to the level of the C3 vertebral body.

Apart from an early bone marrow relapse in June 1977, he made a complete recovery. In particular, there was no evidence of CNS involvement at any time.

He received his last dose of vincristine in May 1979 and completed his chemotherapy by June 1979. The period of cranial irradiation spanned 19 days in April 1977.

In January 1981 he was referred to the neurology clinic with a three month history of progressive painless wasting and weakness of the shoulder girdle muscles. There was marked bilateral winging of the scapulae, left worse than right. The trapezii, rhomboids, supra- and infraspinati, deltoids, teres major and both sternocostal and clavicular heads of the pectoralis major muscles were wasted, more on the left, and power was reduced to grade 4 on the left and 4+ on the right. There was minimal weakness of the left biceps. The triceps muscles were spared as were the distal upper limb muscles and lower limbs. There was questionable weakness of the orbicularis oculi and failure of frontalis to maintain elevation of the eyebrows. Although his face was thin there was no focal wasting or demonstrable weakness of the other facial muscles. There were no sensory symptoms or signs. Tendon reflexes were well preserved and symmetrical. Plantar responses were flexor.

Investigations at this stage including muscle enzymes, thyroid function, cervical spine radiographs, haematological screen and bone marrow were normal. Electromyographic (EMG) studies revealed reduced amplitude ulnar sensory nerve action potentials and evidence of chronic partial denervation of both deltoids, more on the left.

Thereafter the condition appeared to arrest with no objective progression noted during eight years of follow up (1981–9). Serial EMGs showed evidence of chronic partial denervation and reinnervation in the brachioradialis, biceps, deltoids, supraspinatus and trapezius muscles without pathological activity at rest. No significant abnormality was demonstrated in the quadriceps. In the right tibialis anterior a full interference pattern contained occasional polyphasic units of normal amplitude and duration which were not felt to be of clinical significance. Muscles