responded when his name was called, followed simple instructions, and focused objects. The rigidity was markedly reduced enabling the patient to extend his limbs. Grasp and root reflex, pursing reflex and non-habituating glabellar sign disappeared. After 4 months he used up to six words and recognised his relatives.

The patient was admitted to a nursing home and had long-term stabilisation. He died from pneumonia 18 months later, and no autopsy was obtained.

Comment

We report a patient's history of anti-Ta-associated paraneoplastic neurological syndrome (PNS) with an occult testicular malignancy despite unsuspicious tumour diagnostics. Our patient developed atypical parkinsonism with a severe hypokinetic syndrome, striking reduction of verbal output, and massive rigidity, thereby supporting the findings in five patients of a previous report that parkinsonian-hypokinetic features seem to happen more frequently with anti-Ta encephalitis than with other PNS.² Regarding the revised criteria for PNS, the presence of the well-characterised anti-Ta antibody together with the appropriate clinical and imaging findings in our patient fulfils the requirements for definite PNS.³

The decision for orchiectomy without tumour detection was based on (1) life-threatening deterioration, (2) the remarkable number of patients with neurological improvement after tumour resection in cases of anti-Ta-associated encephalitis, (3) the absence of suspicious masses using magnetic resonance, CT, positron emission tomography and endoscopic imaging and (4) the high frequency of testicular cancer in patients with anti-Ta-associated PNS. At the time of presentation, all previously reported cases with anti-Ta encephalitis had testicular neoplasms.³ Up to now, other tumours were found in females to cause Ta-associated paraneoplastic encephalitis.⁴ The underlying mechanisms are thought to consist of tumour proteins triggering an auto-immunological response against the nervous system.³ The first neurological symptoms of our patient preceded tumour diagnosis by about 6 months, which is the median time in the original report on anti-Ta antibodies.³

A special feature of anti-Ta-related paraneoplastic encephalitis is the response to therapy. This could be partly explained by testicular malignancies that respond better to treatment than breast or lung tumours.² After orchiectomy, our patient had improvement in the neurological symptoms including alertness, spontaneous eye opening, following simple instructions, focusing objects and using a few words. Moreover, the rigidity of all extremities was markedly reduced and most primitive reflexes disappeared.

Given that the immunohistochemical staining in this case revealed microscopic tumour despite unsuspicious findings in repeated testicular ultrasonographic examination, the presence of anti-Ta antibodies together with appropriate clinical findings should prompt every effort to find an underlying malignancy. This case underscores the high specificity of anti-Ta antibodies. According to the new diagnostic criteria, six PNS defining reactivities were identified so far,⁵ among which anti-Ta is the most recently identified. One has to keep in mind that failure to make premortem tumour detection is common, also due to possible anti-cancer

effects of the immune response, reducing tumour enlargement and metastasis. This case demonstrates that even resection of healthy-appearing tissue might, in exceptional cases, be necessary and justified, if extensive investigation failed to demonstrate tumour masses.

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Consent was obtained for publication of the patient's details described in this report.

doi: 10.1136/jnnp.2006.101964

Competing interests: None declared.

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Intracranial fluid filled collection and superficial siderosis

Superficial siderosis (SS) of the CNS is caused by repeated haemorrhage into the subarachnoid space with resultant haemosiderin deposition in the subpial layers of the brain and spinal cord.^{1–4} The commonest neurological manifestation is that of slowly progressive gait ataxia and impaired hearing. A remote history of injury is common. A fluid-filled intraspinal cavity of variable dimension is a frequently noted finding on neuroimaging.^{3–6} A high prevalence of CSF containing cysts in SS supports the hypothesis of bleeding from a dural defect as the origin of SS.⁷ Removal of the likely bleeding source is a logical therapeutic strategy.

Case report

A 58-year-old man was evaluated for a 10 year history of progressive decline in hearing and imbalance. Additional symptoms included a 5 year history of slurred speech and a 1 year history of increasing falls. His past history was remarkable for a motor vehicle accident at 7 years of age. This was followed by a prolonged hospitalisation but no evident neurological sequelae. His examination was remarkable for decreased hearing, an ataxic dysarthria, a wide based ataxic gait with increased lower limb tone and hyperreflexia. Rapid alternating movements with the hands and feet were irregular and the heel–shin and finger–nose tests were positive. T2 weighted head MRI showed typical MRI changes of superficial siderosis with the characteristic hypointensity involving the cerebellum (fig 1A), brainstem (fig 1A) and the frontotemporal regions. Also present was post-traumatic encephalomalacia involving the anterior frontal lobes. A CSF study was remarkable for increase in red blood cells to 380 cells/mm³ and a protein concentration of 94 mg/dl. No abnormality was noted on head and neck magnetic resonance angiography. A small cystic lesion was noted behind the cerebellum on T2 weighted axial (fig 1A) and sagittal head MRI (fig 1B), and on axial head CT (fig 1C). This was felt to be of no consequence until a review of the head CT (fig 1C) with careful attention to the bone sequence (fig 1D) revealed discontinuity of the inner skull table underlying the cyst (fig 1D). A CT myelogram of the spine, performed to look for a bleeding source, showed extravasation of the dye from the fourth ventricle into the cystic structure (fig 1E1 and E2).

Discussion

Recent reports have noted that intraspinal fluid filled collections are frequently seen in SS.^{3,7} In some cases the associated dural defect has been repaired surgically, and decrease or resolution of the cavity and CSF red blood cells has been documented.^{3,5,6} This experience prompted us to surgically explore the identified intracranial cystic structure. A burr hole was opened in the superolateral portion of the right suboccipital region. A CSF filled cavity was encountered within the bone. The outer table was removed, a defect in the inner table was identified and was noted to overlie a large dural defect (fig 1F1). CSF from the fourth ventricle was flowing directly out of the defect into the cavity. Nervous tissue discoloured by haemosiderin deposition was noted. A fascia lata graft was harvested from the right thigh and was used to patch the dural defect (fig 1F2). Macerated muscle was placed over the repaired dura which was then buttressed with Gelfoam.

A fluid filled collection in the posterior fossa is often interpreted as an incidental cyst or prominent cisterna magna. The overlying fracture of the inner table and the perioperative finding of a large dural defect communicating with the cyst suggests that the cause was likely post-traumatic. Intraspinal fluid filled cavities associated with SS are a recently recognised finding. This report suggests that an intracranial dural defect may also be the underlying cause of SS. Bleeding from friable vessels in the cyst wall of intraspinal fluid filled cavities has been considered to be a likely bleeding source.³ Our patient had no demonstrable cyst wall. Friable vessels in the diploic space were the

Figure 1 A T2 weighted axial head MRI shows haemosiderin deposition along the cerebellar folia and around the brain stem (single arrows). Also shown is the posteriorly located cystic structure (double arrows). (B) T2 weighted sagittal head MRI shows the posteriorly located cystic structure. (C) Axial head CT showing the cystic structure (single arrow) and fracture of the inner table (double arrow). (D) Axial head CT (bony window) showing the discontinuity in the inner skull table. (E1, E2) CT myelogram showing extravasation of contrast from the fourth ventricle (E1) into the cystic structure (E2). (F1) Dural defect noted at the time of the suboccipital craniotomy. This was leading to the cyst through the defect in the inner table and was overlying the fourth ventricle from which CSF was flowing into the cyst. (F2) Site after repair of dural defect.

likely bleeding source. It is speculative whether exposure of CSF to extradural tissue may lead to SS by an unidentified mechanism. Given the long natural history of SS, the available follow-up duration of 3 months is too brief to assess response to the intervention. Intradiploic CSF fistulas are exceedingly rare.⁸ They are generally benign and are often related to trauma or neurosurgical procedures. No case with associated SS has been reported.

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doi: 10.1136/jnnp.2006.108225

Published Online First 18 December 2006

Competing interests: None.

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PINK1, a gene product of PARK6, accumulates in α -synucleinopathy brains

α -Synucleinopathy is an entity of neurodegenerative diseases such as Parkinson's disease (PD), dementia with Lewy bodies (DLB) and multiple system atrophy (MSA), that involves accumulation of α -synuclein in the brain. PINK1 (PTEN induced kinase 1) is a novel gene recently identified as causative in autosomal recessive early onset parkinsonism (PARK6). In the present study, we examined the localisation of PINK1 in the brains of patients with α -synucleinopathy and found PINK1 in glial cytoplasmic inclusions (GCIs) in MSA, as well as in Lewy bodies (LBs) in PD and DLB. These findings imply that PINK1 may be involved in the formation of LBs and GCIs, suggesting that PINK1 is one of the major pathological proteins in α -synucleinopathy.

Methods

The cDNA of PINK1, corresponding to 112–520 amino acids of the protein, was subcloned in a vector pET30(a) with a His tag. Anti-PINK1 antibody was generated against recombinant His tagged PINK1 by immunising a rabbit. The obtained antibody was affinity purified. A postmortem brain sample from a normal patient was homogenised, subjected to sodium dodecyl sulphate-polyacrylamide gel electrophoresis and transferred to a membrane. After blocking in Tris buffered saline with 5% dry milk, the membrane was incubated with anti-PINK1 antibody (1:1000). The membrane was then incubated with a secondary antibody (1:2500; Amersham, Buckinghamshire, UK), and visualised with an enhanced chemiluminescent substrate (Pierce, Rockford, Illinois, USA). Immunohistochemical analysis was carried out with paraffin embedded midbrain sections from patients with sporadic PD, DLB and MSA, and from normal controls (n = 6, 3, 6 and 6, respectively). First, localisation of PINK1 protein in normal human brain was examined by carrying out double staining of PINK1 (1:500) and cytochrome c (1:1000, mouse monoclonal; Pharmingen, Germany). Sections from patients with PD, DLB and MSA, and from normal controls were immunostained with anti-PINK1 antibody as previously described.¹

Results

Immunoblotting analysis revealed that the anti-PINK1 antibody detected a major band of approximately 50 kDa, corresponding to mature PINK1 protein (PINK1 without a