Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) Associated with Seminoma

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We wish to report a case of CIDP in association with seminoma. A 43-year-old man without significant past medical history developed bilateral lower extremity weakness more than upper extremity weakness about 6 months before presentation. Examination was significant for severe weakness (Medical Research Council, MRC, grade 0–1) of the foot and toe extensors and flexors, moderate weakness of the intrinsic hand muscles (MRC grade 4), mild weakness of proximal lower extremity muscles (MRC grade 4–5) and a steppage gait. Reflexes were hypoactive to absent, and perception of vibration and position sensations were reduced in the distal lower extremities. Laboratory testing included negative tests for HIV-1 and Borrelia burgdorferi infections, absence of monoclonal gammopathy on immunoelectrophoresis, negative antinuclear-antibody (ANA) and a cerebrospinal fluid (CSF) protein of 267 mg/dl (normal range 15–45) with no cells and negative VDRL. Nerve conduction studies are shown in figure 1a. The electromyographic (EMG) needle examination showed scant fibrillations and positive sharp waves and diffusely reduced motor unit action potential recruitment in all muscles examined.

Prednisone 80 mg/day (1 mg/kg) was given for 8 weeks and then slowly tapered to an alternate-day schedule. Three months after presentation a left testicular mass was diagnosed as seminoma and resected, and chemotherapy was started for a left retroperitoneal metastasis. The patient improved prior to the initiation of chemotherapy with normal proximal upper extremity strength and continued weakness of intrinsic hand muscles (MRC grade 3), ankle dorsiflexors (MRC grade 2), plantar flexors (MRC grade 3), and toe flexors and extensors (MRC grade 1). Nerve conduction studies 12 weeks after initiation of prednisone showed improvement (fig. 1b).

The patient continued to improve during the prednisone taper and chemotherapy but unfortunately died of pneumonia while leukopenic; postmortem examination of the nervous system was not done.

The patient described here had an acquired demyelinating neuropathy most consistent with CIDP by clinical and electrophysiological criteria [1]. After this diagnosis was made he was found to have seminoma. It seems plausible that the neuropathy was a remote effect of the tumor. Bird et al. [2] described 3 cases of melanoma with CIDP, 2 of which also demonstrated vitiligo, which is evidence of an immune attack on skin melanocytes. This was taken to imply that the immune attack on the melanoma cells incidentally damaged normal skin melanocytes and also perhaps Schwann cells, which share a common neural crest origin and thus presumably some epitopes with melanocytes. This idea is supported by a report of demyelinating neuropathy as a side effect of injection of melanoma cell preparations.
into patients with melanoma in an attempt to increase the immune response to the tumor [3].

Six prior cases of seminoma associated with neuropathy have been reported [4–9]. In 4 [5–7, 9], the neuropathy improved with treatment of the tumor. One [8] improved with prednisone suggesting an immune mechanism, but was axonal rather than demylinating. Three [5, 6, 9] showed elevated CSF proteins. EMG results in one [8] showed axonal neuropathy and in another [9] both demyelinating and axonal features, but details such as nerve conduction velocities were not given.

The present report is the first to truly document CIDP by EMG findings, although other cases showed features consistent with CIDP as described above. Given that CIDP is an immune-mediated condition, our case may represent the effect of an immune response related to cancer incidentally damaging the nervous system.

References

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Cervical Cord Glioma in an HIV-Positive Patient

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A 46-year-old homosexual man without previous medical history was examined for progressive onset over 1 month of leg tingling, with moderately reduced superficial sensation below T10. Other sensibilities, strength, deep-tendon reflexes, urinary function and fundus examination were normal. His serology for human immunodeficiency virus (HIV) was found positive, and his CD4 count was 250/mm3. The cerebrospinal fluid showed an increased protein level at 0.61 g/l, normal glucose content and 16 lymphocytes/mm3. Spinal cord magnetic resonance (MR) imaging disclosed a fusiform cord enlargement extending from C3 to T3 on T1-weighted and an increased centromedullary signal on T2-weighted sequences. Due to subtle enhancement after gadolinium injection, a tumor was considered unlikely. Cerebral MR imaging was normal. Since blood and cerebrospinal fluid tests were negative for herpes simples virus, varicella-zoster virus, Epstein-Barr virus, and cytomegalovirus, boralia, toxoplasmosis and syphilis, and the clinical course was progressive, an infectious myelitis was considered ruled out. MR imaging findings [1], rapid clinical evolution and CD4 count did not favor vascular myelopathy. Despite receiving a 6-week course of zidovudine (1.5 g/day) and a 2-week course of foscarnet and prednisone 60 mg/day, the patient rapidly worsened. Three months after onset, the patient was tetraplegic, with multi-modal hypoesthesia below C5. MR scans showed an increased size of the intramedullary lesion from C3 to T3, hypointense on the T1-weighted sequence, with a heterogeneous ill-defined contrast enhancement in its central portion (fig. 1a). On the T2-weighted sequence, this segment was diffusely hypointense (fig. 1b). These features were consistent with a tumor. The most frequent spinal cord tumor in AIDS patients is lymphoma, but it usually enhances homogeneously with gadolinium. On the contrary, astrocytomas typically enhance in a heterogeneous and progressive way after gadolinium injection and have indistinct margins. Although rare, low-grade malignancies should be considered when nonenhancing spinal cord tumors are encountered [2]. The patient died few days later from acute respiratory failure apparently directly linked to the cervicomedullary involvement.

Pathological examination revealed a glioblastoma predominant at the C5 level extending up to the medulla (fig. 2). Necrosis was evident within the tumor. A few microglial nodules were seen intermingled. Immunocytochemistry did not show any staining for cytomegalovirus, varicella-zoster virus, Epstein-Barr virus or herpes simplex virus. Very few microglial cells and macrophages were stained with antibodies directed against the HIV antigen p24. No vacuolar myelopathy was observed. Spinal cord gliomas are less frequent than brain gliomas, but have also been described in HIV-positive patients: two spinal cord astrocytomas [3] and, recently, two brainstem astrocytomas have been reported [4]. In our patient, the spinal cord lesion was suspected to be the first manifestation of AIDS and may somehow be related to the HIV infection. The association of glial tumors with HIV infection [3–7], although HIV is a nontransforming lentivirus, raises the question as to their possible relationship, especially in young patients, since the incidence of glial tumors increases with advancing age. Unfortunately, to our knowledge, there is no published data comparing the incidence of brain gliomas between global and HIV populations.

In animals, some have been related to viruses like SV 40. Many events could induce glioma in infected patients. HIV infection of human glial cells in vitro and in vivo has been demonstrated, and HIV could directly induce astrocyte proliferation, partly through increased TNF-α production [8]. Co-infection of HIV with another virus might be an inducer [6]. Immunosuppression might play a role in the development of astrocytomas [9]. HIV induces overexpression of TGF-β, which is immunosuppressant, and could thus contribute to malignant transformation of reactive glial proliferation [10] by enabling neoplastic glial cells to escape immune surveillance.

Glioma must be included in a differential diagnosis of spinal cord lesion in HIV-positive patients, but is rare enough not to be sus-
Second MR study performed 3 months after onset of first symptoms. **a** Sagittal postcontrast T1-weighted sequences (1.5 T; TR 500, TE 20) depict the intraspinal expanding lesion both in cephalad and caudal directions from C3 to T3, with heterogeneous contrast-enhancement predominating at the cervico-thoracic junction from C5 to T1. **b** Sagittal T2-weighted fast-spin-echo sequences (TR 3400, TE 132): cervico-thoracic fusiform enlargement with intraspinal heterogeneous signal.

**Fig. 2.** Microscopy examination at the C5 level. Tumoral infiltration of the spinal tissue: spindle cells with irregularly shaped nuclei and ill-defined cytoplasm were densely packed in fascicles and associated with rare lymphocytes around vessels. HE. ×16.

The fact that antigen p24 was not found in our patient’s lesion does not favor a direct role for HIV, although the infection might have accelerated disease course.

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**References**
Epilepsy, Cysticercosis and Neurocysticercosis in Benin

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Human cysticercosis is a parasitic disease caused by Cysticercus cellulosae, the cestode larva of Taenia solium in pork flesh, which is very prevalent in developing countries, mainly in rural areas [1–6].

In 1992 a case of cerebral cysticercosis (neurocysticercosis) was observed in the University Hospital of Cotonou, Benin, in a sexagenarian from Savalou (Benin) who was cured by praziquantel [7], which launched several investigations.

Savalou’s Investigation

This case initiated a neuroepidemiological, clinical and serological investigation on a representative sample of 1,443 subjects living in Savalou from February 15, to April 14, 1993. Farming occupies 4 of 5 inhabitants in this area. The investigation was performed using a 3-stage cluster sampling method. Sociodemographic and cultural data were collected. 1,443 subjects were interviewed, and 6 ml venous blood were sampled from each participant. Sera were analyzed at a dilution of 1/100 with ELISA using a crude antigen of C. cellulosae.

The diagnosis of epilepsy was based on clinical symptoms (without electroencephalography) and asserted by interview. The cysticercosis diagnosis relies on the presence of either a cysticercus with a pathological examination of a cyst, or typical calcifications on X-rays of the skull and soft parts, or a positive serology result (OD ≥ 0.400).

The neurocysticercosis diagnosis relies on the presence of neurological signs, mainly epileptic episodes and at least one of the cysticercosis diagnosis criteria.

Results

More than half (55.7%) of the surveyed people are 5–29 years old and 66% of them are farmers, mostly animists and Christians. Most of them have no access to latrines and prefer consuming pork, the meat being only slightly cooked and without any veterinary control. Pigs are bred roaming freely.

The clinical prevalence of epilepsy is 1.5% (22 cases) and that of cysticercosis is 3.95% (57 cases), with a positive predictive value of 30.7% and a negative predictive value of 99.3%. In 59.1% of the patients, epileptic episodes are complex, and in 27.3% they are generalized. The seroprevalence in cysticercosis patients is 3% (43 cases), and 25% of the sampled subskin nodules (16/64) really bear cysts of cysticercus [8, 9].

Pilate’s Investigation

From April 18, to May 17, 1994, sera obtained from a representative sample of 319 subjects living in Vêkky (Atlantic Department) were tested for anti-cysticercoid antibodies and showed a seroprevalence of 3.45% [10].

National Investigation

2,625 sera obtained from a representative sample of the whole population of Benin during a neuroepidemiological survey on HTLV-I/II conducted from March 1988 to May 1989 were also tested for cysticercosis. 41 were found to be positive, yielding a prevalence of 1.6% with a significant male predominance.

Comments

As a whole, these results appear to list Benin among the countries of cysticercoid middle endemicy with sacs of hyperendemia at Savalou (3.95%) and at Vêkky (3.45%), corresponding to places of intense pig farming and pork consumption.

In fact, the practice of X-ray of the skull and the soft parts and that of the cerebral tomodensitometry would have contributed to a better estimation of the prevalence of this disease. Risk factors of the disease are encountered in all the areas of the country: pigs roaming free, their slaughtering being without veterinary control and the consumption of meat which is not well cooked. But a large campaign "Information – Education – Communication” has been started in the different areas to educate in pig breeding, hand washing, use of latrines, and in the consumption of well-cooked pork meat. The impact of these prophylactic measures remains to be assessed.

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