P27. Withdrawn

P28.

Metastatic Intradural Extramedullary Carcinoma: Case Report and Review of the Literature

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The patient presented with symptoms and signs of cervical radiculopathy and myelopathy. Myelography and computer axial tomography of the cervical spine and brain demonstrated a solitary discrete tumor nodule in the cervical subarachnoid space. At operation, gross examination of the tumor was consistent with schwannoma. Pathological examination revealed renal cell carcinoma. Postoperative abdominal ultrasound exposed a mass involving the kidney.

A review of the literature yields few case reports of intradural extramedullary spinal metastasis originating from a primary tumor outside the central nervous system. In the reported cases over the past twenty-five years, the most common primary tumor was carcinoma of breast, followed by lung, melanoma and uterus. In most of the reported cases, spinal subarachnoid metastasis represents a tertiary deposit seeded by malignant cells in the CSF from secondary lesions elsewhere in the central nervous system. This theory does not account for the pathology in this case report. Alternative theories of metastatic spread have included extension along perineural lymphatic ducts, transdural invasion, and hematogenous dissemination.

P29.

Regional Nerve Injury Following Intra-arterial Cis-platinum and Radiation Therapy for Bladder Cancer: A Follow-up Study

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Patients treated for invasive bladder cancer with concurrent internal iliac artery Cis-platinum infusion and radiation therapy have been reported to have a high incidence of somatosensory disturbance, occurring in conjunction with treatment (Journal of Clinical Oncology, February, 1989). Of 25 patients treated in such fashion, 11 have been symptomatic. This is a report of a detailed neurologic examination and electrophysiologic study on 10 of these 11 patients and on 4 additional patients, 3 of whom received sequential rather than concurrent treatment.

The sensory deficit appeared to be mainly large fibre and axonal in nature, with the distribution of sensory loss in some patients suggesting a peripheral neuropathy, while in other patients having a more definite dematomal distribution. Seven of 14 patients recovered within 12 months, another 3 were only mildly symptomatic at 20 months, and the remaining 4 had considerable dysaesthesia more than 20 months following therapy. Marked asymmetry was noted in 2 of these patients, both of whom had only ipsilatearal Cis-platinum infusion on at least one occasion.

The total dosage of Cis-platinum, and the total amount of radiotherapy, did not correlate with the development of symptoms or signs.

We postulate a direct neurotoxic effect of Cis-platinum, relating to the intra-arterial infusions.

P30.

Malignant Intravascular Lymphoma: Immunochemical Markers in 3 Cases

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Malignant intravascular lymphoma (neoplastic angioendotheliomatosis) is a rare disorder with pathological features of intraluminal proliferation of atypical mononuclear cells in blood vessels involving multiple organs. The central nervous system and skin show predominant involvement and there is relative sparing of the reticuloendothelial system. We report on 3 patients encountered at our center during the past 4 years with this disorder. Brief clinical summaries are provided with photographs of relevant pathological material. In 2 of 3 patients, the illness was characterized by subacute onset of a progressive dementia with multifocal neurological signs and persistent increase in CSF protein. The third patient presented with progressive myelopathy and at autopsy was found to have extensive involvement of spinal cord and nerve roots. Recent reports support histogenesis from lymphocytes rather than endothelial cells. Immunocytochemical studies on these 3 cases are reviewed.

Neuromuscular

P31.

Late Pseudo-exacerbation of Myasthenia Gravis Due to Ectopic Thymoma Invading Lower Cranial Nerves

G. MACLEAN and A. GUBERMAN (Ottawa, Ontario)

An underlying thymoma, locally invasive in 1/3 of cases, is found in 10% of patients with myasthenia gravis. Ectopic cervical thymomas occasionally occur but rarely extend above the hyoid. We report a case of cervical thymoma invading the jugular foramen and lower cranial nerves initially mistaken for an exacerbation of myasthenia gravis diagnosed eight years previously.

The patient presented at age 57 with typical bulbar, ocular and generalized signs of myasthenia gravis confirmed by electrical studies, edrophonium testing and elevated acetylcholine receptor binding anti-bodies. Her course was stormy but she eventually responded to pyridostigmine, prednisone, plasmapheresis and azathioprine. A thymectomy by "sternal-split" procedure nine months after diagnosis identified only residual thymic tissue.

Seven years after diagnosis a right cervical mass appeared with worsening dysphagia and dysarthria initially attributed to myasthenia. CT showed a large right parapharyngeal mass invading the base of the skull near the jugular foramen. Examination demonstrated a hoarse voice, wasting and fasciculations of the right side of the tongue, right palatal weakness, and wasting and weakness of right trapezius and sternocleidomastoid. The excised mass was identified as thymoma by a panel of 10 pathologists and she received a course of cobalt therapy.

We were unable to find other reports of ectopic thymoma invading cranial nerves in patients with myasthenia gravis. The cause of this patient's dysphagia and dysarthria was readily distinguishable from worsening myasthenia on the basis of physical examination.

P32.

Congenital Muscular Dystrophy Associated with "Leukodystrophy" and Normal Intelligence

N.J. LOWRY and D.G. MUNOZ (Saskatoon, Saskatchewan)

Fukuyama in 1960 first reported the association of central nervous system abnormalities with the occurrence of congenital or early onset of muscular dystrophy. The patients he described had mental retardation, microcephaly and muscular dystrophy. In the 1970's, CTS in Fukuyama's type muscular dystrophy were reported as demonstrating marked lucencies in frontal white matter. Subsequent pathology reports showed that the hypomyelination was a development abnormality and not a degenerative or leukodystrophic process. Brooke in his 1985 book alluded to cases of congenital dystrophy with abnormal white matter on brain CT scans but with normal intelligence. We have recently encountered a brother and sister with congenital muscular dystrophy, I.Q.'s of 115 and 110 and a marked "leukodystrophic" CT scan. We conclude CT's should be performed in all cases of congenital muscular dystrophy