tify primary intracranial neoplasms among the population of Rochester, Minnesota, for the period 1935 through 1990. We ascertained 382 cases, 134 males and 248 females. A comparison of the age-specific incidence rates for the periods 1935 to 1977 and 1978 to 1990 showed no change for glioma or meningioma. The overall age-adjusted average annual incidence rate per 100,000 was 15.5 (95% confidence limits 11.4-17.6) for the total, 12.7 (10.5-14.9) for males, and 14.2 (12.7-15.6) for females. The age- and sex-adjusted inci-
dence rates per 100,000 for different types of brain tumors were 4.1 for glioma, 5.0 for meningioma, and 3.9 for pitu-
itary tumors. For gliomas and meningiomas, the age-specific incidence increased progressively with advancing age. The higher incidence of primary brain tumors in females and the excess of meningioma over glioma were attributable to the inclusion of asymptomatic meningiomas diagnosed initially by neuroimaging or at autopsy. Based on 76 live patients, the prevalence of primary brain tumors in the Rochester population on January 1, 1991, was 85 per 100,000. Pituitary tumors increased from 1.9 per 100,000 in 1935 to 1977 to 3.7 per 100,000 in 1978 to 1990; this trend probably re-
sulted from advances in hormonal assays and neuroimaging.

P129. Cerebral Lymphomatoid Granulomatosis
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Lymphomatoid granulomatosis (LG) is an uncommon lym
phoproliferative disease characterized by perivascular pleo-
orphic cellular infiltration and necrosis. The lung is the us-
al primary site with secondary central nervous system
(CNS) involvement in 20% of cases. Primary cerebral LG is a
rare but potentially treatable disease with protein manifesta-
tions. We will describe 5 cases of cerebral LG, of which 3
had no clinical evidence of disease activity outside the CNS.
The neurological manifestations included encephalopathy
followed by seizures in 2, mass lesions in 1, and an illness
resembling multiple sclerosis in 2. Cerebrospinal fluid (CSF)
protein levels were elevated in 4 of 4 patients, and oligoclonal
bands were positive in 1 of 1 of these. However, only 1 patient
had CSF pleocytosis and CSF cytology was unhelpful. Tran-
scranial lung biopsy was performed in 2 patients (1 of whom
had a normal chest x-ray film) and was diagnostic in both,
obviating the need for cerebral biopsy. Two received radio-
therapy, 2 received chemotherapy, and 1 died without spe-
cific treatment. Four patients died 6 months to 2 years (mean
13 mo) following onset of CNS disease, while the fifth re-
mains alive following therapy at 3 years. The clinical features,
laboratory investigations, neuroimaging, and pathological
findings of cerebral LG will be presented.

P130. Immunological and Pathological Study of a
Patient with Anti-Ri-associated Encephalopathy
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Visalia, CA

A previous report described an autoantibody called anti-Ri
found in serum and CSF of some patients with opsoclonus
ataxia (Lause et al, Ann Neurol 1991;29:241-251). Most of
these patients had breast cancer and the syndrome was thought
to be paraneoplastic. Anti-Ri reacts with neuronal nuclear
proteins that are also expressed in the patient's tumor. We
now report the autopsy findings in 1 of these patients. A
73-year-old woman developed progressive ataxia, nystagmus,
upward gaze limitation, and peripheral neuropathy. She died
3 years later of progressive neurological disease. At autopsy,
tumor was not grossly evident. The brain contained perivas-
cular and interstitial inflammatory infiltrates, particularly in-
volving tegmentum of pons and mesencephalon. Deposits of
immunoglobulin G (IgG) were detected in the cytoplasm and
some nuclei of neurons. Elution of the IgG revealed a high
percentage of anti-Ri IgG. The proportion of anti-Ri
IgG in the IgG extract from various areas of the nervous
system's serum and CSF was determined by quantitative
Western blot analysis. Anti-Ri IgG was ubiquitously ex-
presse in all regions of the brain with particularly strong
reactivity in dorsal mesencephalon. The presence of anti-Ri
antibody in neurons and of inflammatory infiltrates in the
brain (particularly brainstem) supports the hypothesis of an
autoimmune basis for the anti-Ri syndrome and suggests that
the antibody itself may play a role in pathogenesis.

P131. Intracranial and Other Neoplasms in Ollier's
Disease and Maffucci's Syndrome
Theresa M. Cheng, Bahram Mokri, David W. Kimmel,
Joseph Parisi, and Krishnan K. Unni, Rochester, MN

Ollier's disease and particularly Maffucci's syndrome are
known to be associated with certain malignancies. The onco-
genetic basis is unknown. Of 93 patients seen at the Mayo
Clinic with multiple enchondromatosis from 1984 to 1993,
85 patients were found to have Ollier's disease and 10 pa-
tients had Maffucci's syndrome. Primary intracranial neo-
plasms were found in 7 patients with Ollier's disease. Two
patients had gliomas (1 with brainstem glioma, the other with
multicentric brainstem and hemispheric Grade II oligoden-
droglioma); 3 had pituitary adenomas; and 2 had chondromas.
Multiple nonskeletal neoplasms were found in patients with
Maffucci's syndrome, and also Ollier's disease including neu-
romas, adenocarcinomas, adenosarcomas, and ovariian tumors.
Sarcomatous degeneration of the skeletal chondromas was
common. Metastasis of the chondrosarcomas were usually to
the skull base, causing cranial nerve palsies and brainstem com-
pression. Identification of patients with Maffucci's syndrome
and also Ollier's disease is essential since periodic surveil-
ance for malignancies is warranted. In addition, targeted studies
on patients who develop neoplasms will be valuable in the
future for determining the mechanisms of oncogenesis in
these cases, and possible development of treatment modal-
ities.

P132. Neurotoxicity in Patients with Small-Cell
Lung Cancer
A. G. van Oosterhuis, P. Buon, P. Haux, J. J. Jolles,
K. N. Twijnstra, J. Habets, G. P. M. ten Velde,
and A. Twijnstra, Maastricht, The Netherlands

There have been several reports of neuropsychological side
effects of the currently used treatment regimen in patients
with small-cell lung cancer (SCLC), including systemic che-
motherapy with or without locoregional and prophylactic cran-
ial irradiation (PCI). Especially PCI was blamed for its neu-
rotoxicity. Thirty-two consecutive patients with histologically
proven SCLC were tested neuropsychologically before (t0),
after chemotherapy, and after PCI. They were examined on the
15 Words Test, the Stroop Colour Word Test, and the
Trailmaking Test. In the precondition and postcondition,
these patients were compared also with a matched control
group of normals. The SCLC patients were seen at different points
in time by a neuropsychologist. A CT scan of the brain was per-
formed before at t0. Patients with brain metastases were ex-
P133. Metastatic Pilocytic Astrocytoma: A Case Report
Kendra Peterson, H. Brent Clark, Mabel Rohr, and Walter Hall, Minneapolis, MN

Pilocytic astrocytomas are typically benign lesions, frequently completely resected and with an excellent prognosis. We report a patient with a hypothalamic pilocytic astrocytoma that recurred in the cerebellum 6 years after initial therapy. The boy was 12 years old when he first presented with 3 months of headache and personality change, and 1 week of vomiting and visual loss. Computed tomographic scan revealed a large hypothalamic/suprasellar mass with obstructive hydrocephalus. A ventriculoperitoneal shunt was placed, followed by subtotal resection of a pilocytic astrocytoma. CSF revealed atypical cells similar to the tumor. He received focal external beam radiation, 5,400 Gy in 28 fractions to left and right lateral fields. He was left with visual loss and panhypopituitarism that required medical replacement. However, he was able to graduate from a regular high school and live independently. He was without new symptoms when on a follow-up magnetic resonance imaging 6 years later a new enhancing left cerebellar mass was noted, outside the radiation field. In retrospect, a very small abnormality at this site may have been present on the original scan. His examination was notable for no stigma of neurofibromatosis, normal mental status, bilateral optic atrophy, only vision of color and motion, nystagmus, mild left facial weakness, and mild slowing of left fine-finger movements without ataxia. Gross total resection of the well-demarcated mass revealed a pilocytic astrocytoma, similar to the original tumor. This unusual case demonstrates that pilocytic astrocytoma rarely seeds the CSF with the capacity for late and distant recurrence.

POSTER PRESENTATION:
NEUROPHTHALMOLOGY

P134. Association of the 11778 Mitochondrial DNA Mutation and Demyelinating Disease
Kevin M. Flanigan and Donald R. Johns, Baltimore, MD

Leber's hereditary optic neuropathy (LHON), a maternally inherited form of acute visual loss, is most commonly associated with a mitochondrial DNA mutation at nucleotide position 11778. Recently, 6 white women with clinical multiple sclerosis (MS) and 2 white women with demyelinating white matter lesions who had bilateral visual loss and the 11778 mutation were described; no neurological disease was detected in more than 50 men with LHON (Harding, Brain 1992). We present 4 cases (2 white women, 1 black woman, and 1 white man), all of whom had positive oligoclonal bands and MRI abnormalities consistent with demyelinating disease. All had acute visual loss (age at onset ranged from 21-43 yr). Three had other neurological signs and symptoms and were diagnosed with MS. These cases support the possibility of a demyelinating disease and the presence of the 11778 mtDNA mutation and suggest that the association is neither gender nor racially limited.

P135. Visual Phenomena Limited to the Hemianopic Field in Lesions of the Central Visual Pathways
Michael Vaphiades, Gastone G. Celesia, and Mitchell Bregil, Chicago, IL

Patients with homonymous hemifield defects due to ischemic infarction of the visual pathways were studied prospectively. Each patient was tested with Goldmann perimetry, complete neuro-ophthalmological examination, and color vision testing. A detailed questionnaire was used to assess the extent of the patient's symptoms and awareness of the deficit. Spontaneous visual sensations in the blind hemifield were subdivided into (1) phosphenes characterized by unstructured flashes of light or zig-zag lines; (2) photopsias characterized by regular and often repetitive visual patterns (flowers, geometric figures, etc.); (3) visual hallucinations characterized by complex scenes perceived, at least temporarily, as real. Hemianopic agnosia was defined as unawareness of the hemifield loss; hemiachromatopsia was defined as loss of color vision in the spared areas of the affected hemifield; and palinopsia was defined as the persistence of the visual image after the image has been removed. The prevalence of these phenomena was: hemianopic agnosia 25%; hemiachromatopsia 15%; visual hallucinations 30%; photopsias 38%; phosphene 23%; and palinopsia 15%. A correlation between visual phenomena and extent and location of magnetic resonance imaging-verified lesions was obtained. The higher occurrence of these visual phenomena in our study is most likely related to the detailed examination of each patient with a visual field defect.

POSTER PRESENTATION:
NEUROPHARMACOLOGY

P136. Antidystonic Effects of Novel Sigma Ligands

The antidystonic effects of two novel sigma ligands were tested in a rat model of torticollis. A role for sigma receptors in the pathophysiology of dystonia was suggested by (1) abnormal sigma binding in a mutant strain of dystonic rats; (2) the tendency of neuroleptics with sigma activity to produce dystonic reactions in humans; and (3) the ability of unilateral intratruar macrosection of sigma ligands and sigma-active neuroleptics to produce torticollis in rats. Two potent and selective sigma drugs (BD1047 and BD1065) were developed as possible antidystonic agents. In radolignand binding studies, the drugs were found to have a 100-fold or better affinity for sigma sites, as compared to other tested receptors (opiates, PCP, muscarinic, dopamine, alpha-1, alpha-2, 5-HT1, and 5-HT2). For the behavioral studies, chronic indwelling guide cannulae were implanted above the red nucleus. Torticollis was quantified as the angle of deviation of the head after unilateral microsection. In normal animals, BD1047 or BD1065 had no significant effects on head posture. However, these drugs markedly ameliorated pharaco-