

prednisone was unsuccessful.⁹

In view of our patient's excellent response to treatment, we believe that the diagnosis of infiltrative polyneuropathy should be aggressively pursued in patients with hematologic ma-

lignancy and clinically significant peripheral neuropathy of unknown cause. Because this process can be patchy, and because negative sural nerve biopsies have been reported,^{2,14} we believe that biopsy of a clinically

involved sensory nerve is advisable. Empiric treatment for recurrent malignancy may be justified in some cases.

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Horner's Syndrome From Hypothalamic Infarction

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• We report a case of Horner's syndrome due to ipsilateral posterior hypothalamic infarction, occurring in the absence of other signs of hypothalamic dysfunction. Associated symptoms of contralateral faciobrachial weakness and dysarthria correlated with the extension of the infarct into the posterior limb of the internal capsule seen by magnetic resonance imaging. The likely vascular anatomy of this lesion is discussed.

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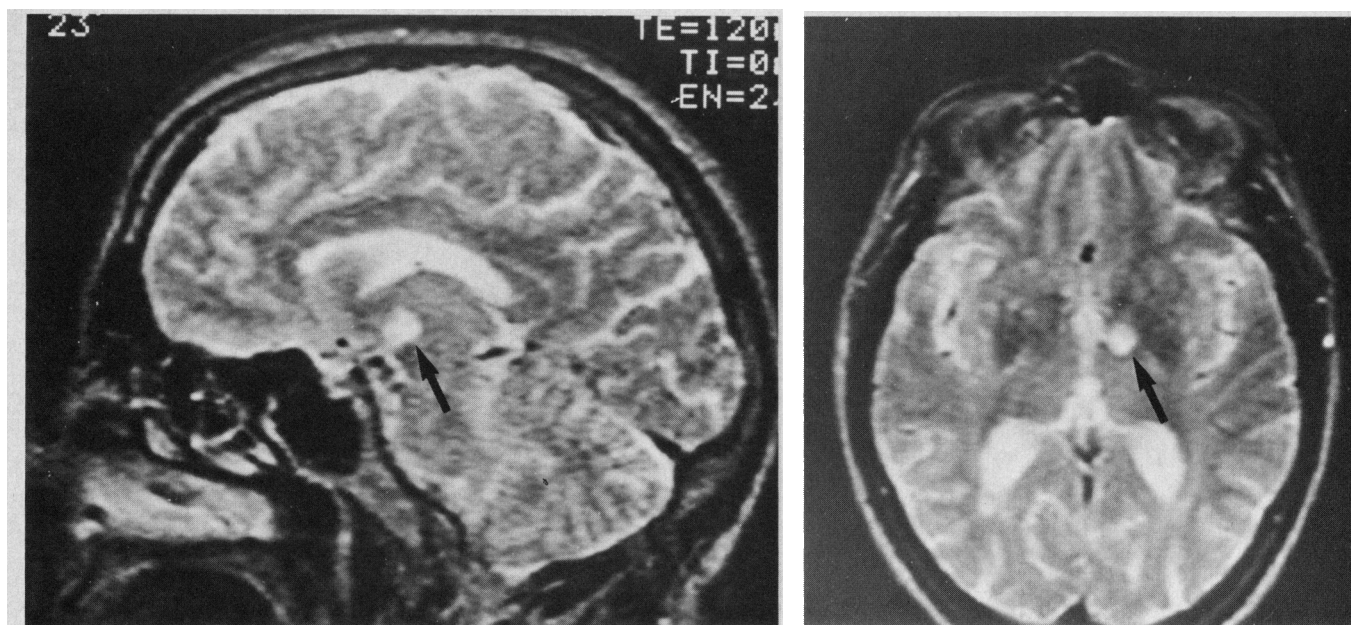
Horner's syndrome can result from interruption of the sympathetic pathway at any point from the hypothalamus to the orbit. The central neuron is involved in as many as 63% of cases,¹⁻³ most of them at the level of the brain stem; few are caused by lesions that implicate the sympathetic cell bodies in the hypothalamus. We are reporting a case of oculosympathetic paresis from focal, ipsilateral, posterior hypothalamic infarction.

REPORT OF A CASE

A 62-year-old woman with hypertension, diabetes mellitus, and coronary artery disease presented with left-sided occipital headache, left-sided ptosis, and right-sided face and arm weakness of 8 hours' duration.

On the day before admission, she was

mildly dysarthric. She was "sleepy" on the day of admission, and, at 6 PM, her left upper eyelid drooped and she complained of left-sided occipital headache. Examination 8 hours later revealed an obese woman with a regular pulse rate of 63 beats per minute, a blood pressure of 135/90 mm Hg, and a crescendo-decrescendo systolic cardiac murmur of grade 3/6. Her mental status was normal, and there was no aphasia. The right pupil measured 4.0 mm and the left 2.0 mm. There was dilatation lag of the left pupil that accentuated the anisocoria when the lights were dimmed, but pupillary reactions were otherwise normal. She had left-sided ptosis, elevation of the left lower eyelid, and decreased sweating over the left side of the forehead. There was slight lingual dysarthria, mild right-sided upper motor neuron facial weakness, right pronator drift, and right arm weakness. Computed tomographic scan of the head, performed



Magnetic resonance images in the sagittal (left) and axial (right) planes (TR = 2000 msec; TE = 120 msec) demonstrating an area of high signal intensity consistent with infarction in the left posterior hypothalamus (arrows).

without contrast, demonstrated a small cavitated infarction in the left corona radiata, lateral to the head of the caudate nucleus. The laboratory test results were as follows: blood glucose level, 19 mmol/L; fasting cholesterol level, 8.25 mmol/L; and triglyceride level, 4.89 mmol/L. An electrocardiogram showed evidence of an old antero-septal myocardial infarction, left ventricular hypertrophy with strain, and first-degree atrioventricular block.

The patient was admitted with a provisional diagnosis of carotid artery dissection, but arteriography gave unremarkable results; a persistent fetal pattern was not present. Neuro-ophthalmologic examination the next day showed normal visual acuity, color vision, and anterior segments. Applanation intraocular pressure was 18 mm Hg bilaterally. The eyelids and pupils were as on admission. Both pupils dilated after instillation of hydroxyamphetamine; however, the next day, neither pupil dilated after 4% cocaine was instilled.

Magnetic resonance imaging of the brain showed an 8 × 10-mm lesion in the left posterior hypothalamus, extending into the genu and posterior limb of the internal capsule. The lesion was slightly hypointense on T₁-weighted images and hyperintense on T₂-weighted images, consistent with recent infarction (Figure). The only other abnormality was the left frontal corona radiata lesion seen on computed tomographic scan. There were no lesions in the brain stem. There were no other signs of hypothalamic dysfunction, and thyroid function and prolactin levels were normal.

The patient was discharged, remained on a regimen of aspirin, and was asymptomatic thereafter. On examination 4 weeks later, her pupils were unremarkable, there was a slight narrowing of the left palpebral fissure, but no face or arm weakness.

COMMENT

Pharmacological tests localized this patient's lesion to the central or preganglionic sympathetic neuron. Although neither pupil dilated with cocaine, the clinical signs of Horner's syndrome were unequivocal and present only on the left pupil; we cannot explain why cocaine failed to dilate the right pupil.

Most cases of hypothalamic Horner's syndrome are caused by large diencephalic tumors,⁴ hemorrhages,⁵ or, less commonly, infarction.^{6,7} In these cases, localization to the hypothalamus is obvious from symptoms and signs that reflect involvement of the thalamus or other adjacent areas. Isolated hypothalamic infarction is an uncommon event owing to the rich vascular supply of this region, eg, anteromedial arteries arising from the anterior cerebral artery and supplying the anterior hypothalamus, posteromedial (thalamostriate) perforators arising from the proximal portion of the posterior cerebral artery at the bifurcation of the basilar artery and supplying the posterior hypothalamus, and short hypothalamic arteries off the posterior communicating artery.⁸ In the case of a persistent fetal circulation pattern, which occurs in up to 30% of individuals,⁹ the blood supply to the hypothalamus can derive solely from the carotid system, and, thus, be more liable to embolic infarction.

The patient's symptoms of faciobrachial weakness, dysarthria, and

drowsiness provide clues to localization of this lesion. Bogousslavsky and Regli¹⁰ reported a case of faciobrachial paresis and dysarthria as an incomplete "psuedo-opercular" syndrome from a small lenticulocapsular hematoma and emphasized that the genu of the internal capsule was the critical site for lesions that produced this clinical picture. Rascol et al¹¹ reported a case, similar to others reported by Fisher,¹² of dysarthria and transitory confusion without paresis, which was attributed to a lesion in the genu of the internal capsule. Infarctions in the distribution of the anterior choroidal artery, which frequently involve the genu and posterior limb, also often cause dysarthria and lethargy.^{13,14} Our patient's magnetic resonance imaging showed that the hypothalamic lesion responsible for the central Horner's syndrome extended into the genu and posterior limb of the internal capsule.

Thus, the symptoms and signs in our case appear to be explainable by involvement of the posterior hypothalamus, as well as the overlying genu and rostral segment of the posterior limb of the internal capsule. Occlusion of a hypothalamic branch of the posterior cerebral or posterior communicating artery is the likeliest cause of the infarction, probably from an embolus of cardiac origin to the posterior circulation. Although lacunar infarcts of the hypothalamus are exceedingly rare (C. M. Fisher, MD, oral communication), given the patient's history of chronic hypertension and the presence of a

small infarction in the frontal white matter, the hypothalamic lesion may also have been lacunar.

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Successful Treatment of Pure Myopathy, Associated With Complex I Deficiency, With Riboflavin and Carnitine

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• We describe a 6-year-old boy who presented with progressive muscle weakness. Additional investigations revealed the existence of a myopathy and a pure motor neuropathy. Biochemical studies in muscle tissue showed a defect of NADH dehydrogenase (complex I). The patient dramatically improved on treatment with riboflavin and L-carnitine. Seven months after the start of the treatment, complex I activity was determined again and appeared to be normalized. Normalization of the enzymatic defect at this level has not been reported before. We provide a survey of nine patients with pure myopathy, associated with complex I deficiency and onset of symptoms in childhood.

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The primary mitochondrial myopathies form a clinically, morphologically, and biochemically heterogeneous group of inborn errors of metabolism affecting the energy-generating pathway.^{1,2} The underlying biochemical defects can be classified into

four groups³ as follows: (1) defects of substrate transport; (2) defects of substrate utilization; (3) defects of the respiratory chain; and (4) defects of energy conservation and transduction.

There is no correlation between the clinical picture and the biochemical defect. Patients with a defect of the respiratory chain may present from early infancy to adulthood. A decreased activity of NADH:Q₁ oxidoreductase (NADH dehydrogenase or complex I) is frequently found. Only nine patients with a combination of symptoms and onset at an infantile age and a clinically pure myopathy, associated with a complex I deficiency, have been described to date.⁴⁻¹⁰ In these patients the diagnosis of complex I deficiency has been established by indirect estimation of NADH-dependent substrate oxidation rates. Effective treatment of mitochondrial myopathies remains extremely limited.

We describe a 6-year-old boy who presented with a progressive myopathy and motor neuropathy in whom complex I activity was found to be deficient. The defect could be demonstrated by indirect estimation of the NADH oxidation rate and by direct measurement of NADH:Q₁ oxidoreductase activity. His condition dramatically improved during treatment with riboflavin and L-carnitine.

REPORT OF A CASE

The patient was the first child of healthy, unrelated parents. Pregnancy, delivery, and motor development were normal. His family history was unremarkable. He had been completely well until August 1987. At that time he started to complain about weakness in his legs, and it was noticed that he was subject to sudden falls. He became unable to run and play games at school. He had difficulty in climbing stairs. Two months later, he developed weakness in his hands to such an extent that he became unable to hold a cup. His gait became waddling and he needed help with his daily activities. Three months after the start of his complaints he was referred to our hospital.

On examination, the patient was an alert and cooperative 6-year-old boy. Results of general physical and ophthalmologic examination revealed no abnormalities. On neurologic examination the cranial nerves were normal. There was a mild weakness of neck flexion, an increased lumbar lordosis, and winging of the scapulae. He had a waddling gait and was unable to sit up from the supine position without using his arms. Gowers' sign was positive. Muscles were generally thin, with a moderate wasting of the proximal musculature. Muscle tone was impaired. There was a diffuse weakness (shoulder girdle, Medical Research Council [1] MRC grade 3; forearms, grade 4; intrinsic hand muscles, grade 3½; pelvic girdle, quadriceps muscle, and hamstring muscle, grade 3; and ankle flexion and extension, grade 4) of all muscle groups that was more pronounced in the proximal musculature.

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