Spread of disease beyond locoregional boundaries is considered extensive disease and is incurable. The majority of cases of localized small-cell cancer of the head and neck presented in the published literature have been treated with local modalities of surgery, radiation, or both in the absence of systemic chemotherapy. Tumors of the major salivary glands are associated with a better prognosis than small-cell carcinomas of the lung or nonsalivary gland sites in the head and neck region, with 2- and 5-year survival rates of 70% and 46%, respectively. Survival approaching 2 years or more has been reported in three patients with small-cell carcinoma metastatic to cervical lymph nodes, without an identifiable primary tumor. However, the median survival of patients with primary small-cell cancers of the larynx, hypopharynx, and trachea is between 7 and 11 months; these tumors seem to demonstrate a more aggressive clinical course. Although fewer in number, more recent case reports and series support the present use of concurrent chemoradiotherapy regimens for limited-stage disease and 5-year survival rates of 70% and 46%, respectively. Survival approaching 2 years or more has been reported in three patients with small-cell carcinoma metastatic to cervical lymph nodes, without an identifiable primary tumor.

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CASE 2. Central Neurogenic Hyperventilation As a Complication of Richter’s Syndrome

A 75-year-old man with a 3-year history of B-cell chronic lymphocytic leukemia (CLL) presented with a reduced performance status and tachypnea for 3 days. He was restless and anxious, but awake and fully oriented. Vital signs were normal except for a respiratory rate of 40/min. Physical examination and routine laboratory tests including blood cell counts, serum glucose, creatinine, lactate, liver function tests, and thyroid-stimulating hormone, did not reveal any abnormal findings besides the features of a typical CLL (Fig 1A). However, arterial blood gases showed the following: pH 7.7; PaO₂, 135 mmHg; PaCO₂, 7.3 mmHg; base deficit, −11.7 mEq/L; and standard bicarbonate, 9.5 mEq/L; consistent with a severe, chronic respiratory alkalosis with incomplete metabolic compensation. In a diagnostic work-up, pulmonary and cardiac causes of hyperventilation were excluded by chest x-ray, ECG, and echocardiography. None of the patient’s drugs was known to be associated with hyperventilation. A psychogenic hyperventilation was excluded, as symptoms persisted during sleep. Cerebral computed tomography scan excluded high intracranial pressure and bleeding. Magnetic resonance imaging (MRI) showed discrete signal alterations within the pons and medulla oblongata on T2-weighted images, but no further evidence for intracerebral mass effect or infarction (Fig 2). Examination of the CSF demonstrated the following: protein, 173 mg/dL; glucose, 9 mg/dL; and leukocytes, 2/mm³. Albumin CSF-serum ratio was 22.6, indicating severe disturbance of the blood-brain barrier. The CSF cytogram showed 6% neutrophils, 22% lymphocytes, 28% monocytes, and 44% large blastoid cells (Fig 1B). The latter cells were CD19+, CD20+, CD5+, immunoglobulin M+, and monoclonal for SK+, corresponding to the known CLL. Microbiological tests were negative. The findings suggested the diagnosis of central neurogenic hyperventilation (CNH) due to CNS infiltration of a transformed CLL. Midazolam, 5 mg intravenously, resulted in immediate reduction of respiratory rate, but the effect was

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Anu Gaba, Rowland Mbaoma, Dwayne Breining, Richard V. Smith, Jonathan J. Beiter, and Missak Haig Went Jr
Albert Einstein Cancer Center, Montefiore Medical Center, Bronx, NY
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Schmid et al

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Transient, and repeated administration was necessary. The patient received intrathecal cytosine-arabinoside, methotrexate and dexamethasone, and intravenous prednisolone. With this treatment, the respiratory rate went down to normal within 4 days. However, the patient developed bilateral peripheral facial nerve palsy, dysarthria, and orofacial dyskinesia that did not respond to high-dose intravenous methotrexate. He subsequently refused any further treatment and died from progressive lymphoma within several weeks. An autopsy was denied by the patient’s family.

This case is unique because of the combination of symptomatic CNS involvement as a rare manifestation of Richter’s syndrome and CNH as an uncommon complication of CNS lymphoma.

Transformation into an aggressive large-cell lymphoma, referred to as Richter’s syndrome, occurs in 3% to 5% of CLL patients. Whereas infiltration of the brain or meninges by typical B-cell CLL is common, only seven cases of Richter’s syndrome with cerebral involvement have been reported.2 CNH was first described in 1959.3 The syndrome is commonly seen in deeply comatose patients with serious CNS injuries, but is a rare finding in patients awake, where it is most frequently caused by an intracerebral malignancy. It is characterized by sustained hyperventilation persisting during sleep, a marked decrease in PaCO₂, elevated PaO₂, and severe respiratory alkalosis. Absence of any respiratory stimulus is mandatory for the diagnosis. So far, 19 cases of patients with tumor-induced CNH have been published, and 10 of these patients suffered from CNS lymphoma.4 To our knowledge, this is the first report on CNH caused by Richter’s syndrome. Several pathogenetic mechanisms have been suggested, including the infiltration of the pons by tumor cells without destruction of the tissue architecture.5 This might lead to medial pontine dysfunction and unopposed stimulation of respiratory centers in the medulla.4 CNS lymphoma are known to develop extensive microscopic infiltration of the brain that may lead to unspecific T2 hyperintensity or may not be detected on MRI.6 In our patient, the presence of large blasts within the CSF and the discrete MRI findings in the pons suggested an infiltration of this area as an explanation for the development of CNH. Symptomatic treatment consists of repeated or continuous intravenous administration of midazolam, propofol or morphium, or mechanical ventilation. In CNH caused by intracerebral lymphoma, intravenous steroids, chemotherapy, and local irradiation were effective for specific therapy.4,7 If lymphoma cells are demonstrated within CSF, intrathecal chemotherapy seems to be helpful. However, the prognosis of CNH is grave, since in the majority of...
published cases, the patients have died within a few months from diagnosis.

Christoph Schmid, Heinz Diem, Karin Herrmann, and Raymond Voltz
Departments of Internal Medicine III, Clinical Chemistry, Radiology, and Neurology, Ludwig Maximilians Universität, Munich, Germany

German Ott
Department of Pathology, Julius Maximilians Universität, Würzburg, Germany

Michael Hallek
Department of Internal Medicine I, Universität Köln, Köln, Germany

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REFERENCES

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CASE 3. AIDS-Related Kaposi’s Sarcoma of the Gastrointestinal Tract

Kaposi’s sarcoma (KS) is a multifocal disease in which involvement of the gastrointestinal tract is frequent, but often asymptomatic. Visceral involvement is usually associated with a poor prognosis. We report the case of a 28-year-old homosexual man who presented with progressively worsening abdominal pain and stools streaked with blood. He had lost 20 pounds of weight in 2 months. Concurrently, he noticed the onset of scattered dark erythematous plaques on his thighs, back and shoulders. A colonoscopy demonstrated segmental, discontinuous erythematous and purple nodules (Fig 1A) with contact bleeding in his rectum, transverse colon, ascending colon, and cecum. Microscopic examination of a mucosal biopsy from one of these lesions showed fascicles of spindle-shaped cells, forming slit-like spaces, within the lamina propria that were infiltrating between colonic glands (Fig 1B). The patient was tested for HIV and found to be positive, with a CD4 T-cell count of 129 cells/mm³. A diagnosis of AIDS-related KS was made.1,2

Infection with human herpes virus 8 (HHV8) is necessary for the development of KS. The long-lasting expression of HHV8 latency genes is important for KS spindle-cell progression. The lesional spindle cells in our patient’s biopsy showed strong nuclear immunoreactivity for the HHV8 LNA-1 antibody (Fig 1C). The origin of the proliferating spindle cells in KS is uncertain, and they are currently believed to be derived from lymphatic endothelium.3 Indeed, the spindle cells in our case stained with the marker D2-40, a novel monoclonal antibody directed toward an epitope on lymphatic endothelium (Fig 1D).

Weei-Yuarn Huang, Liron Pantanowitz, and Bruce J. Dezube
Departments of Pathology and Medicine (Hematology-Oncology Division), Beth Israel Deaconess Medical Center, Harvard Medical School, Boston, MA

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