European Association of NeuroOncology Magazine

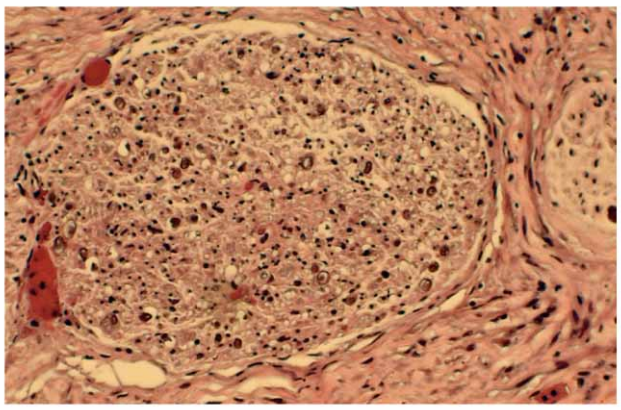
Neurology · Neurosurgery · Medical Oncology · Radiotherapy · Pediatric Neuro-oncology · Neuropathology · Neuroradiology · Neuroimaging · Nursing · Patient Issues

A 39-Year-Old Patient with Double Vision and Rapidly Progressing Bulbar Palsy

Nowosielski M, Glatzer M, Beer R

Maier H, Stockhammer G

European Association of NeuroOncology Magazine 2011; 1 (1) 35-36



Homepage:

www.kup.at/journals/eano/index.html

Online Database Featuring Author, Key Word and Full-Text Search

EANO

The European Association of NeuroOncology

Krause & Pachernegg GmbH · VERLAG für MEDIZIN und WIRTSCHAFT · 3003 Gablitz, Austria

A 39-Year-Old Patient with Double Vision and Rapidly Progressing Bulbar Palsy

Martha Nowosielski[1], Markus Glatzer[1], Ronny Beer[1], Hans Maier[2], Günther Stockhammer[1]

From the Departments of [1]Neurology and [2]Pathology, Medical University of Innsbruck, Austria

Case Report

A 39-year-old male patient with a history of an upper respiratory tract infection 4 weeks before admission presented with headache, vertigo, and abdominal pain persisting for 2 weeks.

Neurologic examination, laboratory findings, gastroscopy, and an abdominal ultrasound remained negative except for leukocytosis (18,100 WBC/µl) and gastrooesophageal reflux disease. Three days after admission to the local hospital the patient developed double vision, dysphagia, blurred speech, and an unsteady gait. Neurologic examination revealed palsy of the sixth nerve, dysarthria, areflexia, and ataxia. Contrast-enhanced cerebral CT showed no abnormalities.

Lumbar puncture (LP) revealed a CSF pleocytosis (90 cells/µl) with protein and glucose within normal limits. A differential diagnosis of encephalitis versus Miller-Fisher syndrome was made. In favour of a viral aetiology the patient received aciclovir 1 g 3×/day and was transferred to the neurology department of our clinic.

Nerve conduction velocity studies and anti-ganglioside antibodies were within normal limits. Cerebral MRI showed diffuse leptomeningeal enhancement (Figure 1). A second LP revealed an elevated CSF protein (61 mg/dl) and pleocytosis (80 cells/µl). Treatment with aciclovir (750 mg 3×/day) was continued and intravenous antibiotics covering listeria spp and legionella spp were added.

Unfortunately, the patient rapidly deteriorated and required artificial ventilation 2 days after admission.

What Is Your Diagnosis?

Meanwhile, the results of the extended CSF diagnostics were available yielding malignant cells in the CSF (Figure 2a) indicating neoplastic meningitis.

In search of the primary tumour, elevated serum CEA (carcinoembryonal antigen 170 µg/l [upper limit 5 µg/l]) and CA 19-9 (carbohydrate 34,815 kU/l [upper limit 37 kU/l]) were detected. A whole-body CT revealed metastases in the

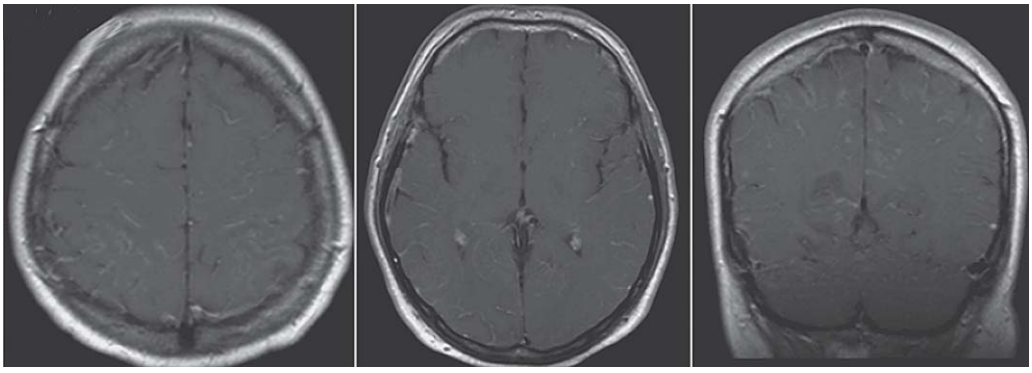


Figure 1. Cerebral MRI (T1 with gadolinium enhancement) showing diffuse leptomeningeal enhancement.

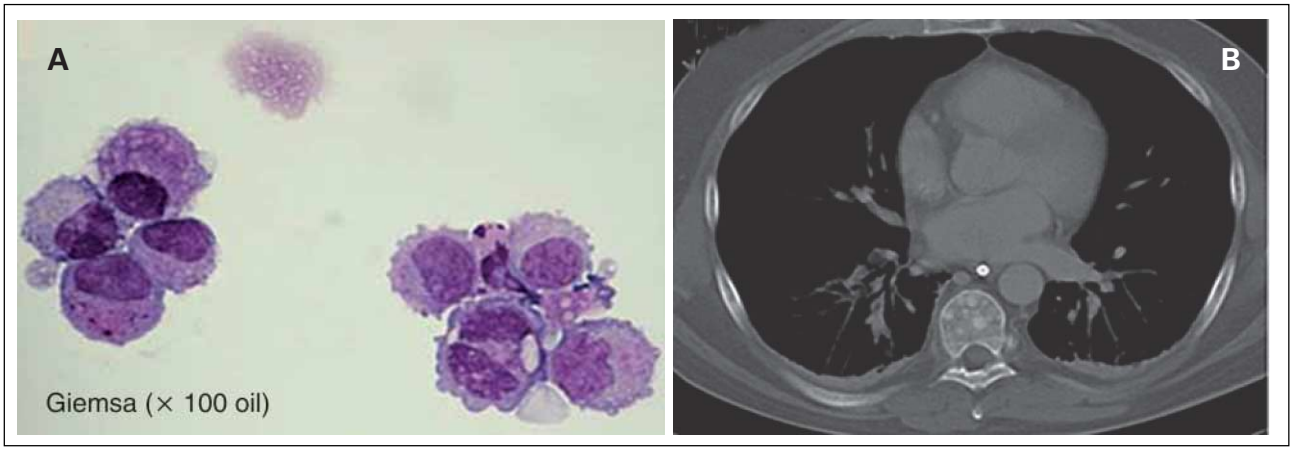


Figure 2. (a) CSF-cytology with clusters of carcinoma cells and **(b)** CT showing bone metastases in the thoracic spine.

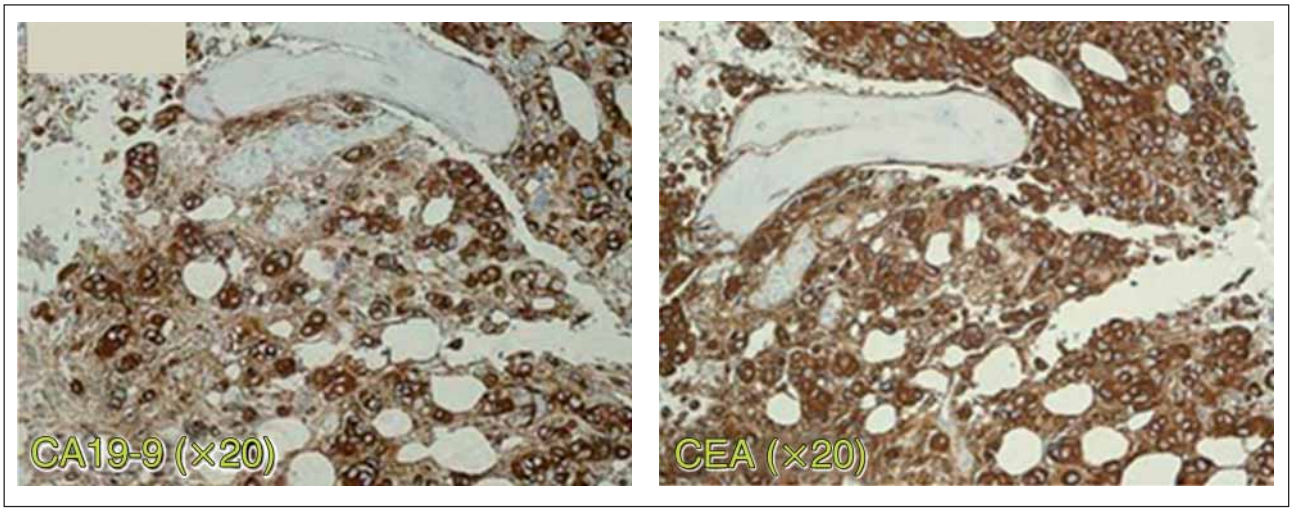


Figure 3. Immunohistochemistry of bone marrow biopsy stained for CA 19-9 and CEA.

thoracic spine (Figure 2b). A bone marrow biopsy showed tumour formations of an anaplastic adenocarcinoma with signet cells and immunohistochemical expression of CEA and CA19-9 indicative of a gastrointestinal malignancy (Figure 3). The clinical course was characterized by a fulminant brain oedema refractory to steroids and other antioedematous therapies and the patient died 5 days after admission to the intensive care unit.

Autopsy confirmed a poorly differentiated gastric adenocarcinoma (Lauren diffuse type, linitis plastica) with peritoneal carcinomatosis, bone metastases, and carcinomatous meningitis (CM) without brain parenchymal metastases.

Comments

This case of CM as the presenting manifestation of gastric cancer is instructional in several respects and deserves some annotation.

CM, which complicates systemic cancers in 3–8 %, represents the primary symptomatic site in up to 5 % of cases with confirmed CM [1]. Although CM was first reported in a patient with gastric cancer, only a few cases have been published in the literature [2]. The largest series of CM complicating gastric cancer has been reported from Korea, where gastric cancer represents the most common malignant solid tumour. In a systematic analysis of 54 cases with CM complicating gastric cancer, CM represented the initial clinical manifestation in 5 of these patients [3].

The patient presented with an unusually fulminant course of CM and gastric cancer was not detected by gastroscopy performed only 2 weeks before the patient's death. Interestingly, a subtype of a poorly differentiated adenocarcinoma called linitis plastica (LP) has been reported to mainly affect young adults and to be particularly aggressive in nature and tricky to diagnose locally [4]. This subtype originates in the submucosa and infiltrates all segments of the gastric wall, resulting in a typically segmental rigidity rather than luminal stenosis [4]. Out of several different possible routes by which cancer cells can reach the CSF space, direct infiltration into the paravertebral venous plexus (Batson's venous plexus) or spinal bone metastases may favour development of CM in this particular type of malignancy.

In conclusion, CM as the initial manifestation of gastric cancer is unusual with 11 patients reported in the literature including our case [5]. Gastric linitis adenocarcinoma represents a particularly aggressive subtype and CM may develop early in the course of the metastatic disease.

Grant Support

MN holds a DOC-fFORTE Fellowship from the Austrian Academy of Science at the Department of Neurology, Innsbruck Medical University.

References:

1. Posner J. Leptomeningeal metastases. In: Posner JB (ed). Neurologic Complications of Cancer. F. A. Davis Company, Philadelphia, PA, 1995; 143–68.

2. Lee JL, Kang YK, Kim TW, et al. Leptomeningeal carcinomatosis in gastric cancer. J Neurooncol 2004; 66: 167–74.

3. Oh SY, Lee SJ, Lee J, et al. Gastric leptomeningeal carcinomatosis: Multi-center retrospective analysis of 54 cases. World J Gastroenterol 2009; 15: 5086–90.

4. Delaunoit T, Boige V, Belloc J, et al. Gastric linitis adenocarcinoma and carcinomatous meningitis: an infrequent but aggressive association – report of four cases. Ann Oncol 2001; 12: 869–71.

5. Braeuninger S, Mawrin C, Malfertheiner P, et al. Gastric adenocarcinoma with leptomeningeal carcinomatosis as the presenting manifestation: an autopsy case report. Eur J Gastroenterol Hepatol 2005; 17: 577–9.

Correspondence to:
Martha Nowosielski, MD
Department of Neurology
Medical University of Innsbruck
Anichstraße 35
6020 Innsbruck, Austria
e-mail: martha.nowosielski@i-med.ac.at