

Acute confusional state in multiple myeloma: consider hyperammonemic encephalopathy

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OBJECTIVES

Hyperammonemia can cause an acute confusional state in patients with multiple myeloma despite normal liver function

CASE REPORT

We present a case of a 77 year-old woman who came to our attention for rapid cognitive impairment developed in the last week, with psychomotor slowness, apathy, echolalia, and postural instability.

In the first days of admission the drowsiness worsened and she became somnolent and unresponsive.

Past medical history was significant for hypertension and recent finding of anemia of unknown etiology.

RESULTS

laboratory findings showed pancytopenia, increased serum level of IgG 24,3 g/L (n.v. 7-16) and kappa free light chain. Blood sodium, calcium, glucose levels and liver function tests were normal. No toxins (benzodiazepines) were detected in the blood.

Blood ammonia level was 68 umol/L (n.v. 11-35 umol/L).

Cerebrospinal fluid analysis was normal. Cerebral MRI scan with gadolinium was normal.

EEG showed bihemispheric slowing, with bilateral periodic triphasic waves, consistent with metabolic encephalopathy (**Fig. 1**)

A bone marrow biopsy confirmed the diagnosis of multiple myeloma.

After a brief course of intravenous dexamethasone the patient showed improvement of vigilance, motor performance and EEG pattern (**Fig. 2**) along with normalization of serum ammonia levels.

At the follow-up visit at three months the patient is undergoing chemotherapy and she is stable. The EEG pattern is unvaried.

DISCUSSION AND CONCLUSIONS

The pathophysiology of elevated serum ammonia in patients with multiple myeloma is not clear. An overproduction of ammonia or an alteration of urea metabolism mediated by the plasma cells have been suggested as underlying mechanisms.

However, the treatment of multiple myeloma (steroids, chemotherapy), has been associated with clinical improvement of the neurological condition.

Given the reversibility of the encephalopathy after adequate treatment, hyperammonemia should be ruled out in case of acute mental state alteration in patients with multiple myeloma.

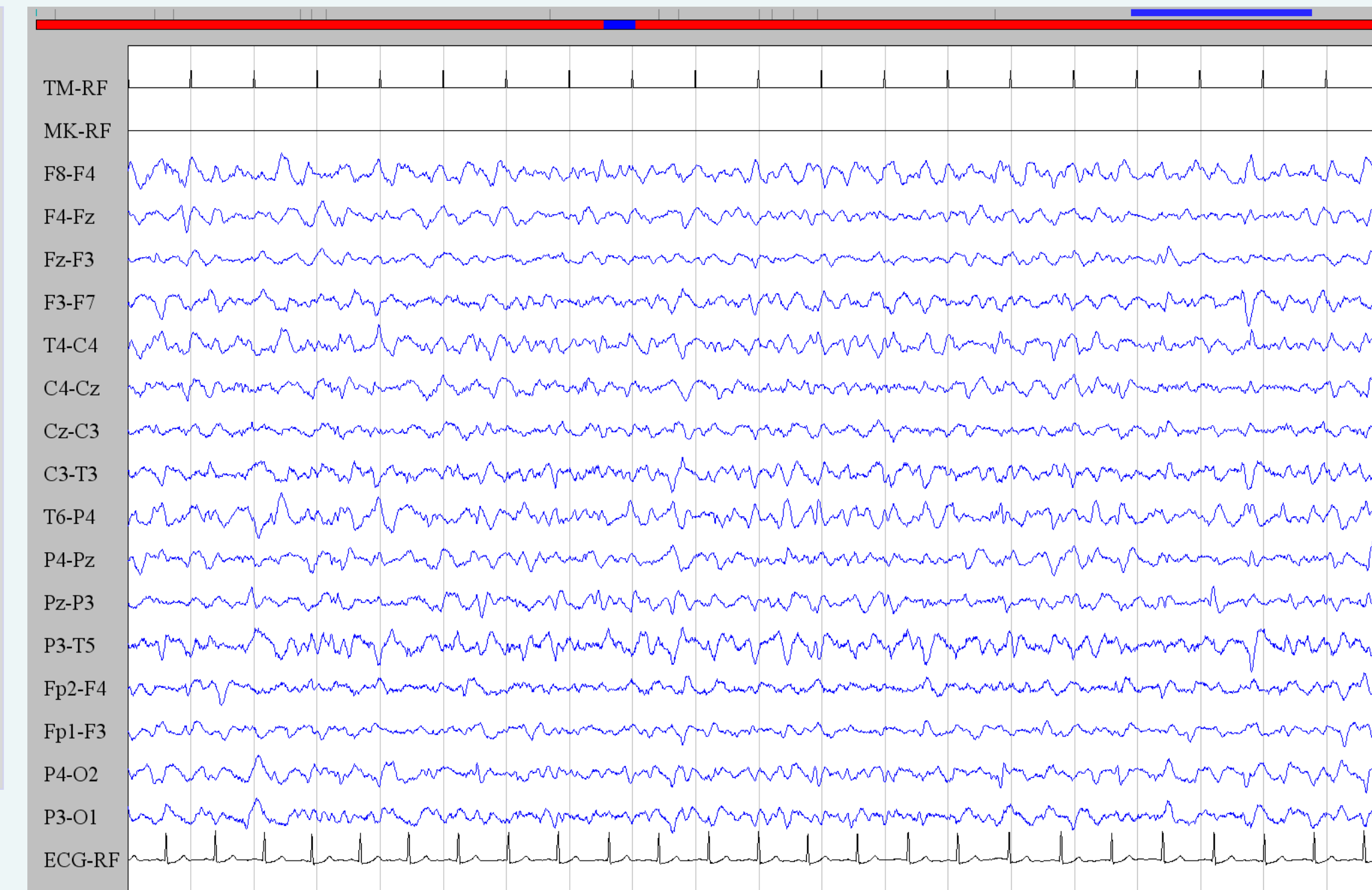


Fig. 1: EEG pattern at first admission

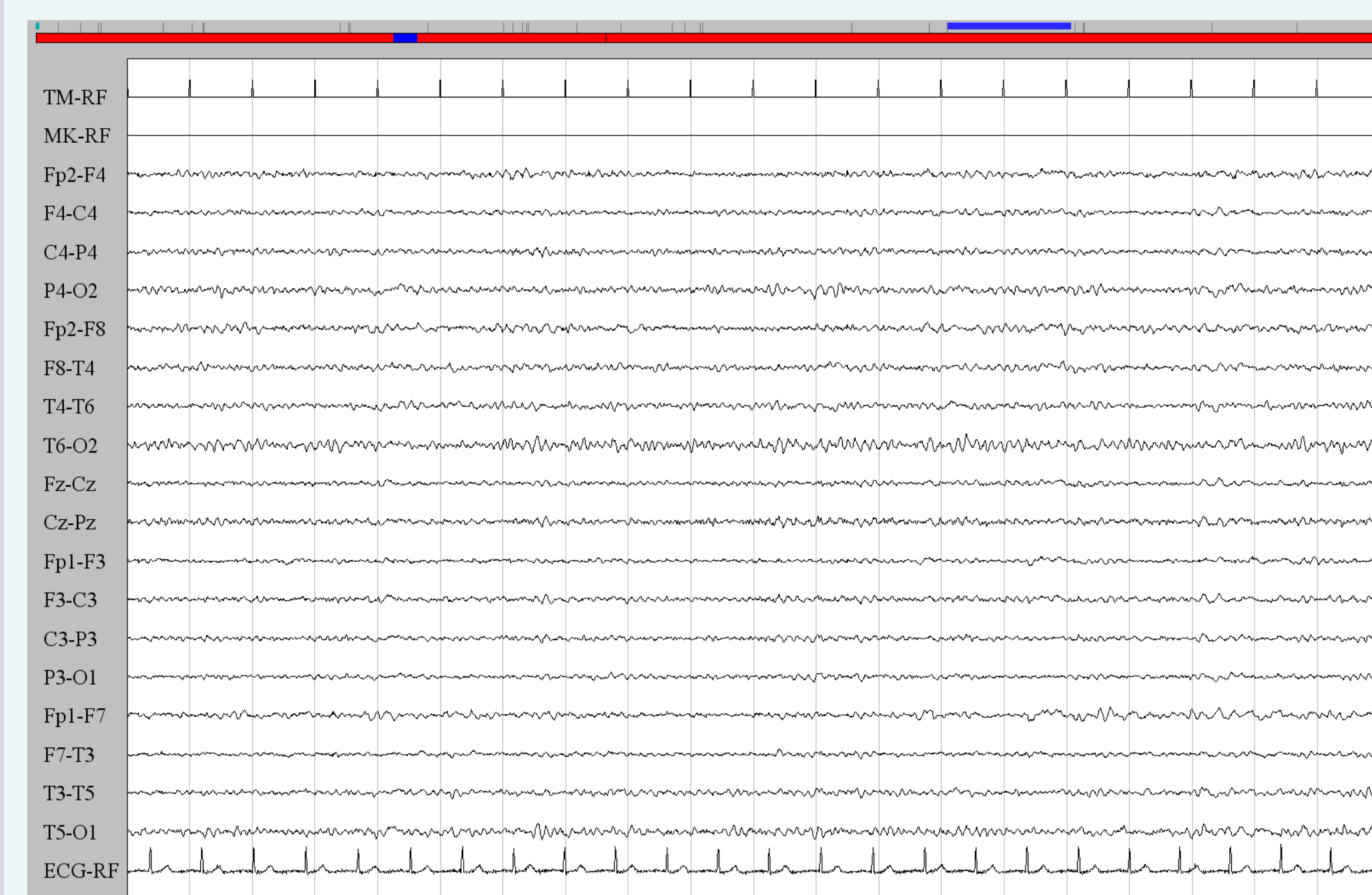


Fig. 1: EEG after one week of dexamethasone