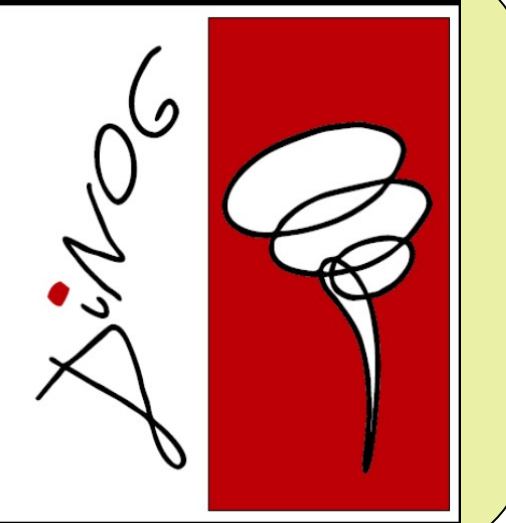




# Acute onset of anti-Hu paraneoplastic limbic encephalitis responsive to Rituximab



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Paraneoplastic limbic encephalitis (PLE) typically presents with a subacute clinical onset and progression of symptoms within weeks to months. According to previous published series, only two cases of anti-Hu PLE symptoms with acute onset have been reported.

Despite aggressive immunosuppressive treatment, the neurological outcome of anti-Hu PLE is poor and only one case responsive to treatment with Rituximab is described in literature.

Herein, we report a case presenting with acute onset of anti-Hu PLE responsive to Rituximab

## CLINICAL DATA Male, 69 years

- 2007 diagnosis of colic cancer, treated with CTX followed by surgical resection
- 2008 diagnosis of prostate cancer, treated with radical prostatectomy
- On July 2012, he was admitted to the Neurologic Unit for acute onset of confusional state and speech impairment, without fever

## NEUROLOGICAL EXAMINATION

- alert but not oriented in space and in time
- MMSE score was 17/30, characterized by severe deficit of short and long term memory (both verbal and spatial), moderate attention and verbal fluency impairment.
- no meningeal or cranial nerves signs were present
- modified Rankin Scale (mRS):5

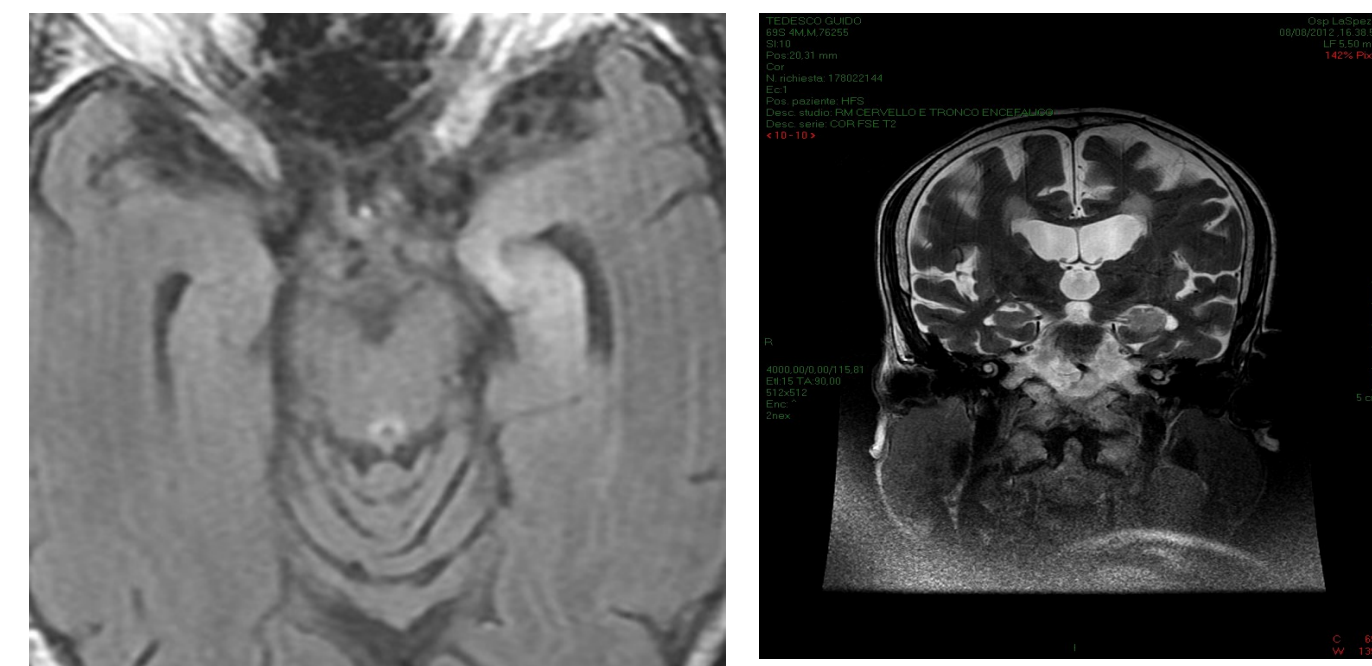
## CSF ANALYSIS

- pleocytosis (40 cells/mm<sup>3</sup>; lymphocytes: 90%)
- increased protein (600 mg/dL)
- oligoclonal bands (only on CSF)
- HSV I-II, VZV, CMV, EBV, HHV6 DNA-PCR: negative

## EEG



## BRAIN MRI



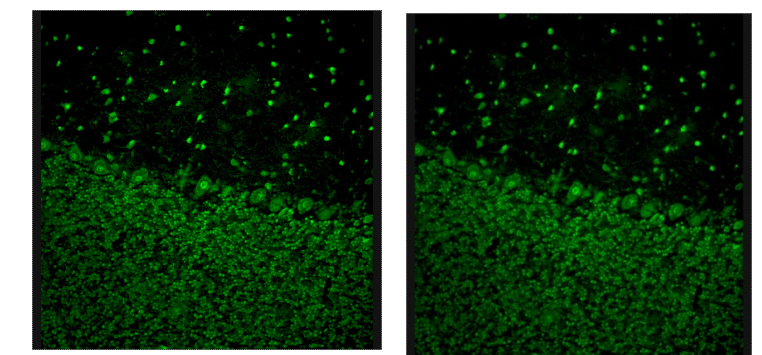
enhancing T2/FLAIR hyperintensity in medial temporal lobes, particularly in amygdalo-hippocampal area, with a moderate swelling and slight diffusion-restricted signal on DWI sequences.

## BLOOD TESTS

Unremarkable, except for:

NSE: 16.2 ng/ml (v.n 0-14.70)

+ Ab anti-Hu



Anti-Hu (ANNA1)

antigen: HuD, HuC, He 1-N1, He 1-N2

## Paraneoplastic Limbic Encephalitis?

TOTAL BODY TC (JULY 2012): unremarkable  
TOTAL BODY PET (AUGUST 2012): unremarkable

Methylprednisolone 1 gr i.v + IVIg 0.4g/kg/die for 5 days

no improvement

**RITUXIMAB** 375 mg/m<sup>2</sup>/week for 4 weeks

## FOLLOW-UP

TOTAL BODY TC (February 2013)

...in the retropharyngeal-perijugular space, a mixed density mass (7x3.5 cm), probably referable to necrotic lymph node... biopsy

**METASTASIS OF NEUROENDOCRINE CARCINOMA**

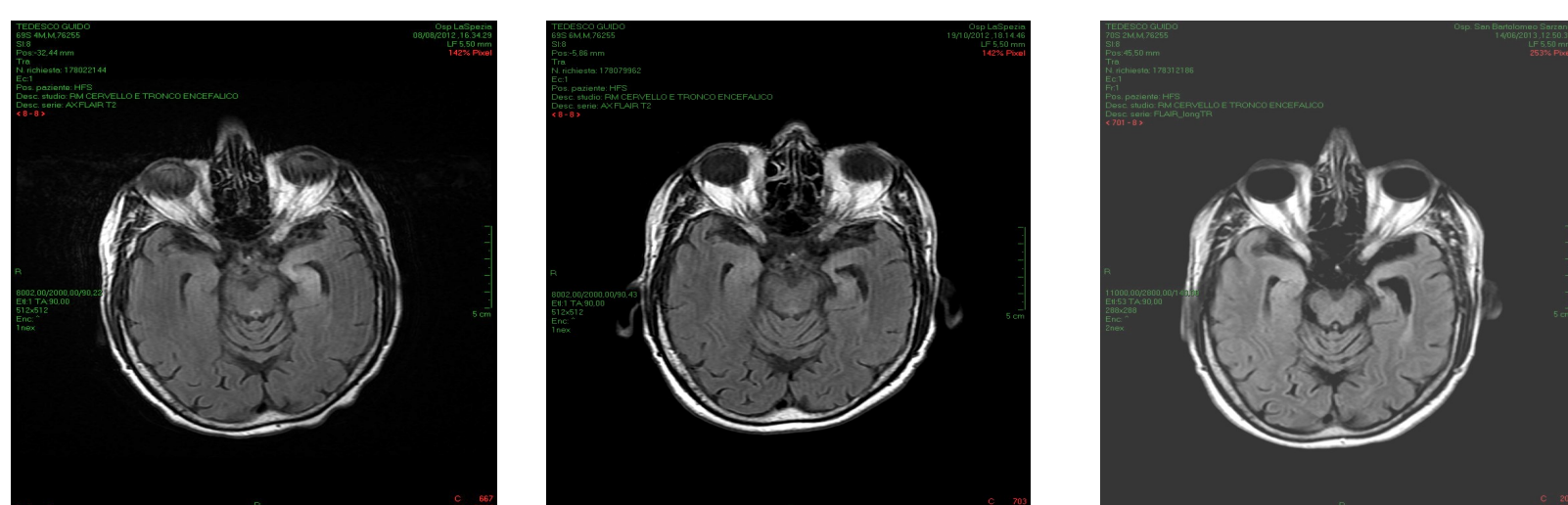
## CHEMIOOTHERAPY

- Cisplatin + Vepesid q21 (III cycles)
- September 2013: Paclitaxel (I cycle)

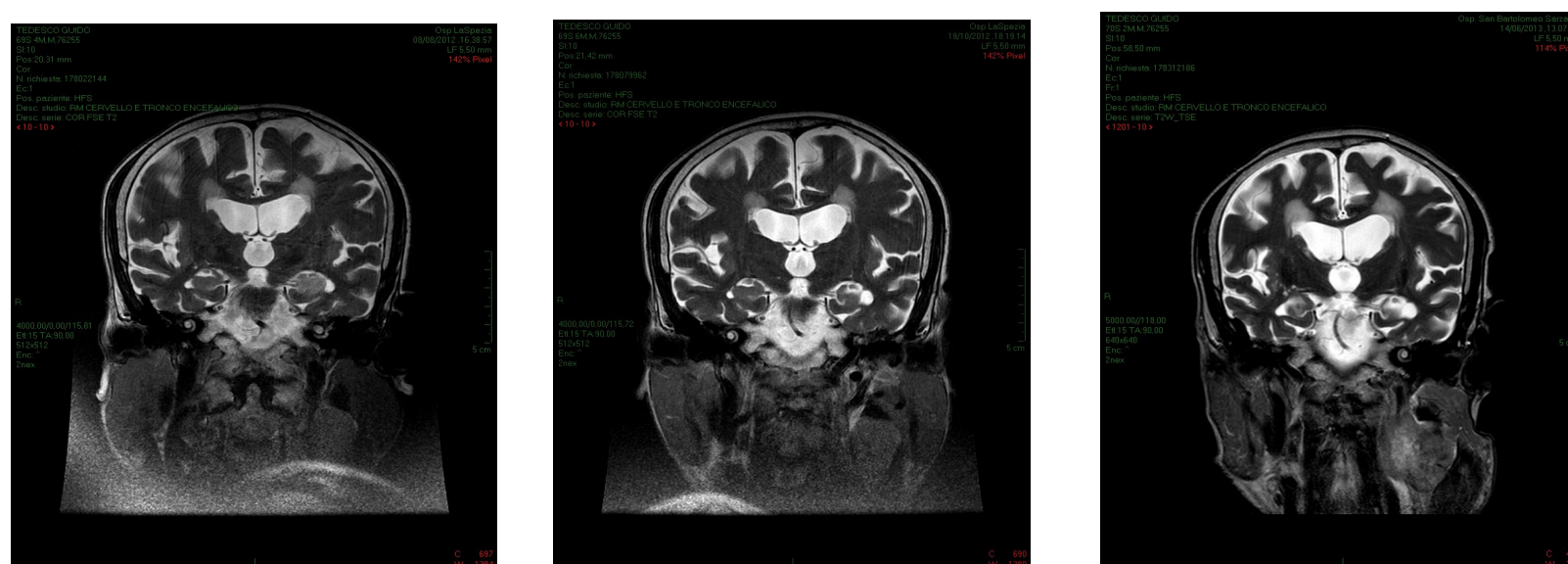
increase in size of the tumoral mass  
severe hyponatremia caused by SIADH

November 2013: exitus

## BRAIN MRI



a) 08-08-12      b) 19-10-12      c) 14-06-13



a: axial FLAIR image shows left mesial temporal hyperintensity as at the first exam

b: axial FLAIR image shows marked decreases of left mesial temporal hyperintensity after two months

c: axial FLAIR image shows mesial temporal atrophy at follow-up MRI examination after ten months

improvement in orientation, memory and verbal fluency

- MMSE score 24/30
- mRS:3 (change RS:-2)

This is the third case of anti-Hu PLE presenting as acute encephalitis. Up to date, only two similar cases are described in literature.

Particularly, despite the aggressive oncological progression leading to death of the patient, a good clinical and neuroradiological response to Rituximab has been observed

increasing of 7 points in MMSE score and reduction of 2 points in mRS

decreasing of mesial temporal hyperintensity, even if with development of consensual atrophy

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