A rare case of neuroendocrine lung cancer in a patient with anti-Hu antibodies

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Background

Anti-Hu antibodies (anti-Hu Ab) belong to the Elav family of RNA-binding proteins. They are usually associated with paraneoplastic neurological syndromes (PNS). In particular, they are considered a tumoral marker for small cell lung cancer, more rarely for neuroblastoma or neuroendocrine neoplasm. PNS show a heterogeneous clinical pattern characterized by bradykinesia, weakness, ataxia, headache, seizures, confusion, personality changes and dementia. In most cases, neurological symptoms precede the discovery of the tumor and resolve when tumor is removed.

Case report

A 55 year-old man presented a few months before with ataxia, numbness, dysesthesias, weakness, cramps and myalgia. **Anti-Hu Ab high titer was found in the serum** and confirmed by immunofluorescence analysis. In the suspect of PNS, we performed:

- tumoral markers (NSE, Cyfra 21.1, α-fetoprotein, CA 125, CA 15-3, CA 19-9, CEA, PSA): negative
- chest X-ray: negative
- cerebrospinal fluid analysis: proteins, cells and IgG were increased
- neurophysiological exam: mild demyelinating-axonal motor and sensitive polineuropathy
- brain MRI: unremarkable
- total body computed tomography: negative.

Endovenous immunoglobulins and plasma exchange were attempted without benefit. Several months thereafter symptoms worsened, with inability to walk unaided, hallucinations, confusion, disorientation, large involuntary movements in the arms and facial grimaces.

Neurological examination showed: nystagmus, anisocoria (dx>sn), dysphagia, dysarthria, dysmetria and severe pseudo-athetosis movements and ballism in the upper limbs, mainly on the right.

Electroencephalogram was slow and disorganized.

Brain RMI was repeated and revealed signal abnormalities of left anterior amygdala and hippocampus (Fig, 1). So the patient was treated with high dose of e.v. steroid therapy, with significant improvement of involuntary movements and cognitive functions, followed by oral Azathioprine 100 mg/day.

Total-body FDG-PET uncovered intense metabolism in left supraclavicular node, histologically compatible with metastasis from lung NE (TTF1+, SINAPTOFISINA+, CK7+).

Octreo-Scan with Octreotide-In111 confirmed the presence of a **lung neuroendocrine carcinoma at left inferior lobe**. Chemotherapy with Cisplatin was then started.

Conclusion

We aimed to underline the unusual clinical presentation of PNS related to neuroendocrine lung cancer in this patient, who manifested a cerebellar syndrome followed by severe limbic system involvement and to suggest to screen also for neuroendocrine lung cancer in the presence of anti-Hu Ab.

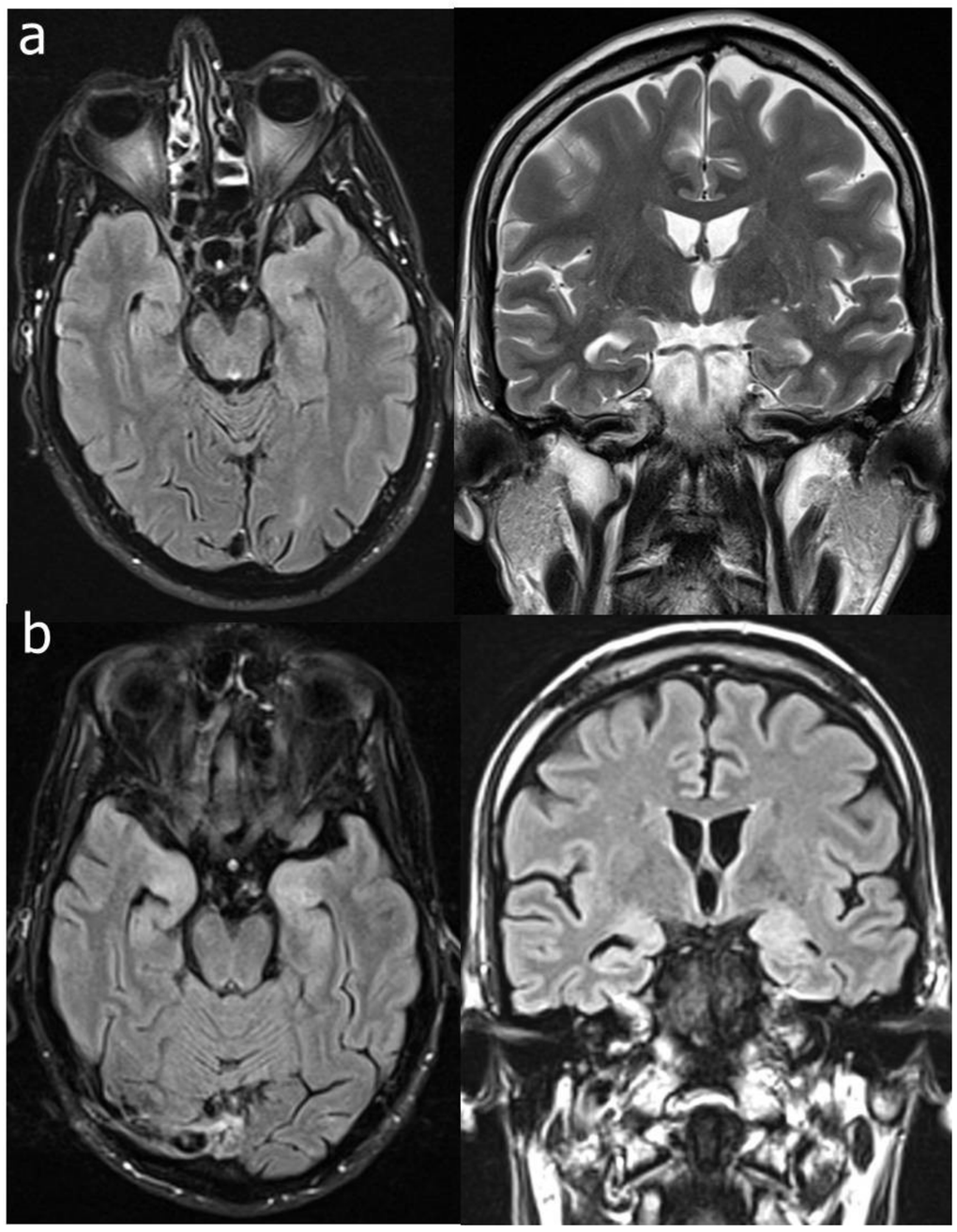
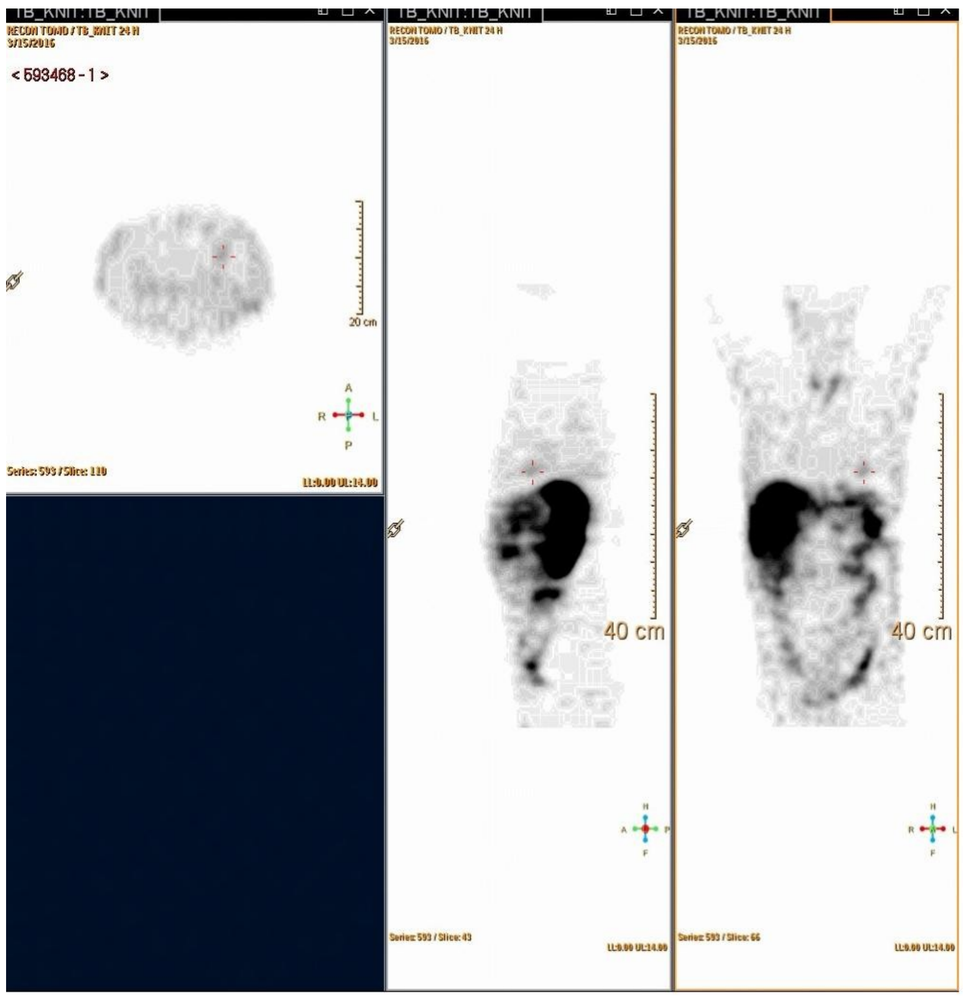


Fig.1. Brain MRI images were normal at disease onset (fig. a) but showed signal alterations in both mesial anterior temporal lobes at eight-month follow-up (fig. b).



Octreo-Scan with Octreotide-In111: lung neuroendocrine carcinoma at left inferior lobe.

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