Available online at www.elixirpublishers.com (Elixir International Journal)

Physiology and Anatomy

Elixir Physio. & Anatomy 110 (2017) 48418-48419

ISSN: 2229-712X

Paraneoplastic Autoimmune Encephalitis Associated With Testicular Cancer- About One Case and Literature Review.

Hicham Ouazize, Tony Saad and Nizar Badran
Centre Hospitalier Sud Essonne Dourdan-Etampes France.

ARTICLE INFO

Article history:
Received: 11 August 2017;
Received in revised form:
7 September 2017;
Accepted: 18 September 2017;

Keywords
Limbic encephalitis,
Paraneoplastic disorder,
Testicular cancer,
Testicular microlithiasis.

ABSTRACT
Paraneoplastic limbic encephalitis is a rare clinical entity. It is clinically manifested by neuropsychiatric disorders that accompanies or precedes most often the discovery of the cancer.We report an interesting case of paraneoplastic limbic encephalitis with anti-Ma2 antibody in a man of 51yers with only apparently benign microcalcification of the testes on imaging, and whose histopathological examination revealed a seminomatous germ cell tumor.

Introduction

Paraneoplastic limbic encephalitis associated with testicular cancer is a very rare condition, usually in a subacute setting. It manifests clinically in a neuropsychiatric way that most often accompanies or precedes the discovery of cancer. Secondary paraneoplastic limbic encephalitis due to a testicular cancer, comes second after bronchial carcinoma 80% of cases, other cancers can be seen: the breast; thymus, lymphoma. [1]

Observation

We report a case of a 51-year-old patient with a history of a type 2 diabetes and high blood pressure. The patient has been experiencing a decrease in activities with a generalized fatigue syndrome, hypersomnia of anterograde amnesia and disorders of higher function. The evolution has been characterized by the onset of a generalized tonic-clonic convulsive crisis. A cerebral CT scan was performed showing diffuse cortico-subcortical atrophy associated with left ventricular and temporal hypodensities without further signs (Fig. 1), the lumbar puncture was discretely haemorrhagic but recovered a hyperprotéinorachie at 0.99g / l and hypercellularity at 16 elements (mainly PN).

An etiological assessment has been performed with realization of negative HIV / hepatitis C serology. PCV CMV, ABV, enterovirus, lyme, VZV, EBV, mycoplasma and negative syphilis. The autoimmune balance is also negative, in the CSF the antibodies anti-HU, Anti-Yo, Anti-Ri, Anti-CV2, Anti Amphysine are negative, On the other hand the anti MA2 is strongly positive.

The presentation is therefore in favor of a paraneoplastic limbic encephalitis with anti-neuronal MA2 strongly positive in the CSF, parallel to the search for a paraneoplastic origin has led us realized a CT-CAP which does not find an anomaly, the markers LDH , alpha Foteo protein, HCG, PSA in norms, testicular ultrasound finds many hyperechoic images evoking calcifications (stage 3) of the left testicle but without visible mass (Figure 2).

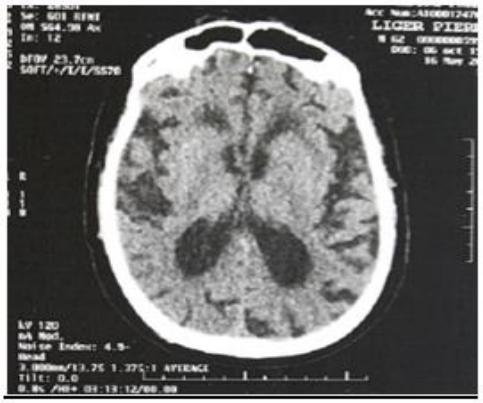

Figure 1. CT image showing diffuse cortico-subcortical atrophy associated with left peri-ventricular and temporal hypodensities.

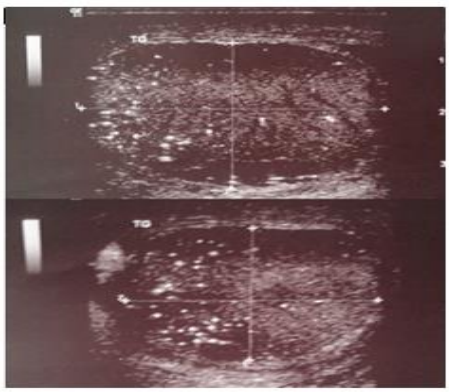

Figure 2. Testicular ultrasound showing numerous hyperechoic images suggestive of calcifications (stage 3) of the left testis but without visible mass.

The patient was presented and discussed in multidisciplinary consultation meeting, and in view of the absence of IVIg improvement, a left orchiectomy was performed which was performed inguinal and histological examination in favor of a seminomatous germ cell tumor).

Tele:
E-mail address: h.ouazize@gmail.com

The evolution was marked by a marked improvement in neurological status after several weeks.

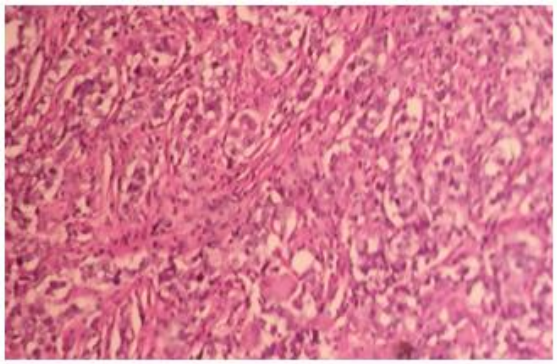

Figure. 3. Tumor proliferation arranged in diffuse layers separated by fibrous septa thickened with some lymphoid islands, the cells are polygonal with clear cytoplasm with rather sharp boundaries, the nuclei are hyperchromatic which in favor of a seminoma.

Discussion:

Paraneoplastic limbic encephalitis is a rare, usually subacute, condition. It most often accompanies or precedes the discovery of a cancer [2].

The pathogenesis of this disease is not completely determined, but the most retained hypothesis is that of an autoimmune reaction against the antigen coexpressed by tumor cells and neurons causing neuronal loss and also a lymphocytic and microglial infiltration perivascular system [3].

The clinical symptomatology of paraneoplastic limbic encephalitis is a syndrome that includes a change in personality, irritability, depression, convulsions, memory deficiency, dementia [4]. These symptoms are due to dysfunction of the limbic system (hippocampus, amygdala, hypothalamus, and insular and coredulate Cortex) [5]. Neurological signs of extralimbic involvement can be seen in 42% of cases, including more diffuse cerebral involvement, cerebellar ataxia or peripheral neuropathy [6].

The CSF analysis contributes to the diagnosis by demonstrating the absence of a malignant cell to eliminate leptomeningeal metastases with the absence of meningeal MRI contrast. It can show inflammatory syndrome in 64% of cases. The EEG shows epileptic activities in the temporal lobes in 50% of cases, but which remain aspecific [6]. It appears that anti-Ma2 antineurone antibody is strongly associated with testicular cancer but also has been reported in other cancers; bronchial cancer, breast, thymus, lymphoma [1].

The diagnosis of paraneoplastic limbic encephalitis requires histological confirmation or the presence of the following 4 criteria [1]:

1) characteristic clinical presentation,

2) interval of less than 4 years between onset of neurological symptoms and diagnosis of tumor ,

3) elimination of other neuro-oncological complications,

4) at least one of the following paraclinic elements: inflammatory syndrome without CSF malignant cells, temporal signal abnormalities with temporal epileptic activity in EEG. In our observation, these criteria are rounded up.

The association between germinal testicular tumor (GTT) and testicular microcalcifications (TMC) was strongly suspected, a GTT was diagnosed in 0 to 8% of patients with TMC [7-8]. There is no official recommendation regarding the value of TMC. There is a doubt about their association with GTTs and the medical-economic impact probably contributes to the lack of recommendation. Management of testicular microlithiasis requires follow-up with an annual ultrasound examination.

Conclusion:

Paraneoplastic limbic encephalitis is a rare condition, and the prognosis and prognosis depend on those of the primary tumor. The present case highlights a clinical and ethical dilemma regarding the role of orchidectomy as a salvage potential with the problem of obtaining informed consent from a semi-comatose patient.

References:

1. GULTEKIN SH, ROSENFELD MR, VOLTZ R, EICHEN J, POSNER JB, DALMAU J. Paraneoplastic limbic encephalitis: neurological symptoms, immunological findings and tumour association in 50 patients. Brain 2000; 123: 1481-94.

2. Voltz R, Gultekin SH, Rosenfeld MR, et al. A serologic marker of paraneoplastic limbic and brain-stem encephalitis in patients with testicular cancer. N Engl J Med 1999;) 340(23):1788–95.

3. Darnell RB. Onconeural antigens and the paraneoplastic neurologic disorders: at the intersection of cancer, immunity, and the brain. Proc Natl Acad Sci U S A 1996;93:4529-36.

4. Corsellis JA, Goldberg GJ, Norton AR. "Limbic encephalitis" and its association with carcinoma. Brain 1968;91(3):481–96.)

5. Bakheit AMO, Kennedy PGE, Behan PO. Paraneoplastic limbic encephalitis: clinico-pathological correlations. J Neurol Neurosurg Psychiatry 1990;53:1084-8.)

6. LAWN ND, Westmoreland BF, KIELY MJ, LENNON VA VERNINOS S. Clinical, magnetic resonance imaging, and electroencephalographic findins in paraneoplastic limbic encephalitis. Mayo Clin Proc 2003; 78: 1363-1368

7. Peterson AC, Bauman JM, Light DE, McMann LP, Costabile RA. The prevalence of testicular microlithiasis in an asymptomatic population of men 18 to 35 years old. J Urol 2001;166:2061-4.

8. Middleton WD, Teefey SA, Santillan CS. Testicular microlithiasis: prospective analysis of prevalence and associated tumor. Radiology 2002;224:425-8.