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Case Report

Cerebral Localization of Waldenstrom's Macroglobulinemia: Bing Neel Syndrome a Case Report

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Abstract

In Waldenström's disease, direct involvement of the nervous system by tumor infiltration is rare and exceptional, forming Bing-Neel syndrome, which groups together the central neurological damage of this disease.

It is associated with neurological manifestations with confusion, temporo-spatial disorientation or motor impairment.

The bing and Néel syndrome must be differentiated and discussed in the face of affections of the central nervous system such as multiform glioblastomas or multifocal leukoencephalopathies as well as primary brain lymphoma.

Keywords: dysglobulinemia; bing-neel syndrome; waldenström; monoclonal immunoglobulin

Introduction

Waldenström's macroglobulinemia (WM) is, according to the WHO classification, a lymphoplasmacytic lymphoma in which the bone marrow is infiltrated by clonal lymphoplasmatic cells associated with M immunoglobulin (Ig) [1].

The cerebral localization of **Waldenstrom's dysglobelinemia** is extremely rare. it results from the infiltration of the central nervous system by malignant lymphocytes. In 1936 BING AND NEEL described a syndrome made of association with hyperglobulinemia; signs of CNS involvement and infiltration of the brain by malignant lymphocytes. Since BING-NEEL syndrome is used to define rare cases of localization of Waldenstrom's disease in the central nervous system.

Case report

Reporting to us the case report of an 80-year-old patient who was admitted to our department for management of hydrocephalus in a double cerebral location.

The patient is known and monitored for WALDENSTROM disease in the hematology department. The treatment followed at home is based on polychemotherapy with good clinical and biological improvement.

The patient is hospitalized at our department for consciousness disorders

On examination we find:

- an unconscious patient (GCS = 8/15)
- the pupils are reactive
- left hemiparesis

As part of the exploration report; a cerebral CT scan performed in him found a left temporal tumor process probably intra ventricular. (Fig 1)

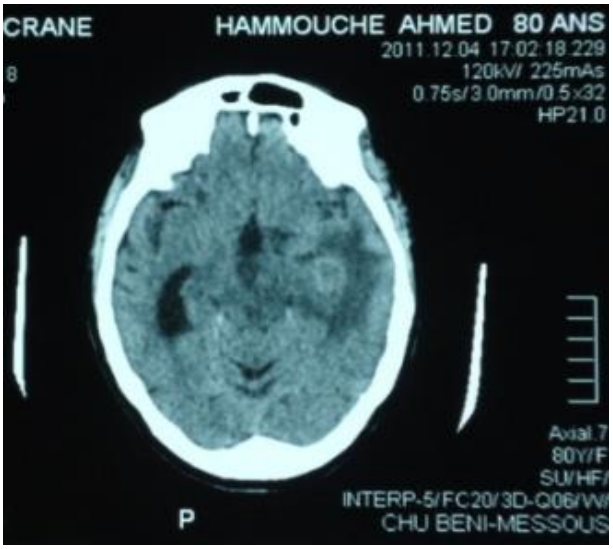

Figure 1: *CT image of a deep tumor nodule*

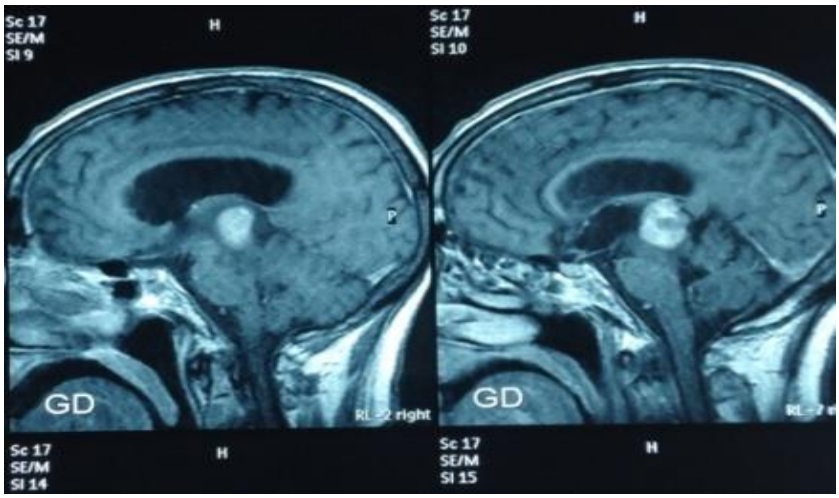

Figure 2: *MRI appearance of the tumor nodule taking the contrast*

Brain MRI revealed a nodular process straddling the post wall of the third ventricle and TECTUM, obliterating the midbrain aqueduct and responsible for active upstream hydrocephalus (Fig 2).

A second nodule of the same nature in projection of the left hippocampus measuring 20mm (Fig 3)

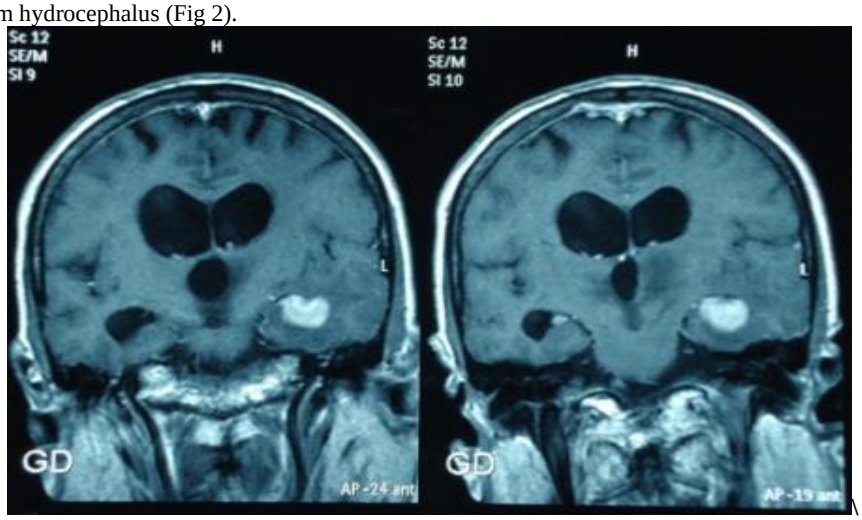

Figure 3: *Appearance of the 2nd tumor nodule in the left hippocampus*

A secondary nature is mentioned first. Faced with the disturbance of consciousness, the patient received a frontal puncture as part of the emergency.

The course is marked by the onset of an epileptic seizure in the upper right limb which became generalized secondarily.

The indication of VCS (ventriculo-cisternostomy) was discussed in our patient in order to treat both hydrocephalus and to perform biopsies of tumor nodules.

CSF immunoelectrophoretic analysis for IGM peaks in CSF was performed. The results confirm the presence of a monoclonal component in the gamma position at a concentration equal to 9.84 g / l CM already typed. This is a lambda light chain IGM.

The course is marked by the death of the patient following complications of his disease.

Discussion

Waldenstrom's disease is a rare condition. It represents 2% of monoclonal gammopathies [2]. Neurological complications are seen in 25% of patients with the disease. The most frequent manifestation is poly sensitivomotor neuropathy.

BING AND NEEL syndrome is the neurological complication of WALDENSTROM disease. It is due to lympho-plasma cell infiltration and deposition of IGM in the brain. It is an extremely rare complication [3].

The clinical presentations of the syndrome are varied, and the diagnosis is often difficult. BING-NEEL syndrome exists in 02 forms [4].

- The infiltrating and diffuse form

- The tumor form

In the diffuse form: lymphoid infiltration which is diffuse to the nervous system which may be expressed by signs of diffuse encephalopathy or signs of meningeal and meningo-radicular involvement)

In the tumor form: it corresponds to an intracranial expansive process which can be single or multi-focal and generally is localized in hemispherical regions under deep cortical. The most frequently encountered clinical signs are intracranial hypertension, focal deficit, partial or generalized seizures [4].

On brain CT and MRI, cerebral localization appears in the form of contrast enhancement and / or thickening in the meninges.

To raise the diagnosis of BING AND NEEL syndrome, in the first place, it is necessary to confirm WALDENSTROM'S disease. Cytological analysis of the CSF, supplemented by immunostaining on a slide, is essential. cortico-meningeal or cerebral biopsy is sometimes necessary for confirmation of the diagnosis in 15% of cases, and MRI is essential to exclude vascular symptoms linked to hyperviscosity, in particular using diffusion sequences (in case of hyperviscosity-related ischemic disease) [5].

Conclusion

BING AND NEEL syndrome must be differentiated and discussed in the face of central nervous system affections such as multiform glioblastomas or multifocal leukoencephalopathies as well as primary brain lymphoma.

The cerebral location of Waldenstrom's disease must be taken into consideration and discussed with pathologies of the central nervous system, especially in a patient with this condition despite its rarity.

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