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A HUGE BING-NEEL TUMOR MIMICKING MENINGIOMA AND REVEALING A WALDENSTRÖM'S MACROGLOBULINEMIA: A CASE REPORT

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Title:

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Key clinical message

Bing-Neel syndrome is a very rare condition referable to an infiltration of the central nervous system by tumoral cells during Waldenström's macroglobulinemia and commonly appears as small size lesions. We report this exceptional case of a Bing-Neel tumor mimicking a huge intracranial meningioma as well as its successful management.

KEY WORDS: Waldenström's macroglobulinemia, Bing-Neel syndrome, lymphoplamatic cells, lymphoma, diffuse, Tumor, meningioma, case report

Manuscript body

Introduction

Waldenström's macroglobulinemia (VM) is a low-grade B cell lymphoproliferative disorder characterized by a monoclonal synthesis and secretion of immunoglobulin M (Ig M) [1, 2]. Neurologic symptoms are commonly a part of hyperviscosity syndrome or autoimmunologic processes [1, 4]. Rarely, they may result from a direct infiltration of the central nervous system (CNS) by lymphoplasmatoid cells and thus defines the Bing-Neel syndrome (BNS) [1, 2, 3, 4]. This CNS metastasis is commonly a diffuse spreading and a focal tumor like presentation is very rare with usually small size lesions [2, 3 4]. An appearance of a genuine extra axial tumor is very rare in the literature and to the best of our knowledge only 3 cases has been found. We report an incidental discovery of a huge fronto-parietal lesion mimicking a meningioma and referable to a Bing-Neel syndrome after pathological and biological investigations.

Case history/ examination

A 56-year-old man was admitted after an incidental discovery of a fronto parietal lesion on a craniofacial computed tomography Scan (CT scan) performed for the investigation of a mandibular tumefaction. His past medical history was remarkable for pulmonary tuberculosis treated 25 years ago. Clinical examination was uneventful apart from a left mandibular tumefaction.

Methods (Diagnosis, investigations and treatment)

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Diagnosis and investigation: Cerebral Magnetic resonance imaging (MRI) revealed a left fronto-parietal extra-axial lesion isointense on T1 weighted images with mass effect and a narrow perilesional edema on FLAIR images. This process measured 9×6.5×4 cm and was intensively enhanced on post gadolinium T1 weighted images with a typical ''dural tail sign" mimicking a convexity meningioma. Moreover, the mandibular tumefaction was identified as a voluminous lymph node. Intra operative findings showed an aggressive tumor with a poor plane of cleavage from the adjacent brain parenchyma. A total resection was achieved and the patient developed a postoperative aphasia and a right hemiparesis, which progressively resolved nearly completely (figure 1). Surprisingly, the first pathological examination revealed a normal lymph node parenchyma and we failed to find such case in previous literature reports. However, additional serological examination revealed an Ig M monoclonal gammopathy (rate of Ig M=29.2g/l) with Lamda light chain rate of 4.12g/l. Erythrocyte sedimentation rate was elevated to 50mm within the first hours. Moreover, Bence-Jones protein in urine was positive with also Lamda light chain monoclonal gammopathy. Protein electrophoresis in the cerebro spinal fluid (CSF) was normal albeit lymphoplamatic cells greater than 5/mm^3 were found. Pathological reexamination of new serial slices of the surgical specimen revealed a lymph node parenchyma which architecture is completely dislocated by a tumoral proliferation consisting of an infiltration of small cells (lymphocytes) with some areas of plasmacytic differentiation associated to few immunoblasts. Some residual germinal center was noticed. Immunohistochemically, the vast majority of the cell population shows a diffuse expression for CD20 and CD79a without expression for CD5, CD10 and CD23. Ki 67 proliferation index was 5%. Therefore, the histopathological and immunohistochemical examination led to the final diagnosis of lympho-plasmacytic lymphoma. A complementary bone marrow biopsy demonstrated a lymphoplasmacytic infiltrate with a similar immunohistochemical profile (figure 2). These pathological and biological examinations were consistent with a fronto parietal Bing-Neel tumor complicating a WM.

Treatment: The patient was sent to the department of Clinical hematology to follow an adjuvant chemotherapy.

Conclusion and results (Outcome and follow-up)

He was asymptomatic at the eighteenth month follow up without tumoral recurrent on the cerebral CT scan.

Discussion

WM is a rare hematologic malignancy accounting for 1-2% of monoclonal gammopathy and

associated to lymphoplasmacytic lymphoma in the WHO classification [9, 10]. This B cell lymphoproliferative disorder is defined by the infiltration of lymphoplasmacytic cells into bone marrow and the demonstration of immunoglobulin M monoclonal gammopathy [4, 8]. Neurologic symptoms occurring in 25% of cases are commonly related to serum hyperviscosity or autoimmunologic phenomena [8]. However, a malignant infiltration of the CNS by lymphoplasmacytic cells remains extremely rare and defines Bing-Neel syndrome. Two Danish physicians namely Jens Bing and Axel Valdemar Neel first demonstrated this exceptional condition in 1936 [7, 8]. According to a literature review, there was only 33 cases of BNS until 2010 on Pubmed. Simon et al reported 44 cases of BNS in a multicenter study and gave a literature review of 33 cases from 1995 to 2014 [11]. However, this last decade (2015 to 2024) showed a great interest for BNS With 105 cases found on Pubmed research out of 154 reported. so far. BNS consists in either diffuse or tumoral form as in our case [1, 2, 4, 7, 8]. Tumoral BNS is often consistent either with intraparenchymal process suggestive for glioma, or meningeal infiltration [8, 9]. Thus, extra axial appearance is very rare and, to the best of our knowledge, only 3 cases of BNS mimicking meningioma are reported in the literature and our case displays the biggest size [6]. The clinical presentation of the tumoral BNS is similar to other space occupying lesions and usually consists of focal deficit or seizure [1, 2, 8, 9, 10]. Diffuse BNS often presents with headache, memory loss, confusion, dementia or coma [1, 4, 8, 10]. In our case, the patient was paucisymptomatic despite the large size of the tumor and let foretell a long duration of evolution. BNS can occurred at any time during the course of the disease [12]. In a study of 34 cases of BNS, Castillo et al noticed this occurrence in half of WM cases within 5 years and in 18% over 10 years [12]. Fintelmann et al classified BNS into 2 categories: group A describes patients with neurologic symptoms probably as a result of lymphoplasmacytoid cells within the

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central nervous system (parenchyma, meninges, CSF and dura); Group B refers to BNS cases with neurological symptoms without cells (< 5 cells/mm^3) within the CSF [13, 14]. Our patient may be classified as group A and some authors suggested a more important damage of blood-brain-barrier allowing the transfer of these cells from the blood to the central nervous system in this group [13, 14]. The diagnosis of tumoral BNS is based on the pathological examination of the tumor and the bone marrow biopsy associated to protein electrophoresis [1, 2, 6]. This pathological examination often demonstrates atypical lymphoid cells consisting of small lymphocytes, lymphoplasmacytic cells and plasma cells [1, 2]. Immunohistochemically, these BNS cells are B cells usually positive to CD20, CD138 and negative to CD5, CD10, CD23 [2, 7, 10]. These cells express immunoglobulin M and serum proteins electrophoresis always shows an Ig M monoclonal gammopathy [1, 2, 8]. All those pathological and proteins immunoelectrophoresis features were found in our patient. The authenticity of our presentation is highlighted by the radiological presentation. Indeed, all but three of the Bing-Neel tumors were intra-axial lesions [6, 8]. The first extra-axial tumoral BNS mimicking meningioma was reported by Civit et al in 1997 [3], the second and third was reported in 2020 and 2023. Our observation is the fourth and We failed to find a most voluminous than this latter in literature review. Because of their intra-axial location and their relatively small size, some Bing-Neel tumor requires only stereotaxic biopsy for histological confirmation [9]. In our case, the radiological diagnosis of the lesion was consistent with a giant meningioma, thus the surgical approach was justified. The treatment of tumoral BNS is based on chemotherapy, which might be associated to a focal radiation therapy [1, 2, 6]. WM shows an estimated median survival of 7-12 years, albeit BNS prognosis was still severe with a free survival rate of 12 months (0-84 months) despite a heavy therapeutic armamentarium [14, 6]. However, the outcome of tumoral BNS seems to be better than diffuse BNS [6] and more recent study revealed a survival of 5 years and 10 years of 71% and 59% [11, 14]. Castillo et al identified in univariate analysis, age above 65 years, previous treatment for WM and platelet count < 100 x 10^9/L as adverse prognostic factors [12]. None of these latter were found in our patient and he was asymptomatic a eighteenth month follow up.

Conclusion

Bing-Neel syndrome remains a very rare complication of WM above all the tumoral form [1, 2, 6, 8]. This uncommon entity might sometimes reveal the monoclonal gammopathy like in our case [1, 6]. Thus, Bing-Neel tumor should be included in the differential diagnosis of not only intra-axial but also extra-axial tumors such as meningioma. The Prognosis improve in time with some long-term survival more than 10 years after successful treatment [14].

Authorship Contribution

Author 1: Ibrahim DAO (corresponding author): Conceptualization of the original idea and writing – original draft.

Author 3: Hassan BAALLAL: Conceptualization of the original idea and contribution to the final version of the manuscript

Author 3: Ousmane OUATTARA: Conceptualization of the original idea and contribution to the final version of the manuscript

Author 4: Narcisse Wendpuiré Mike OUEDRAOGO: Conceptualization of the original idea and contribution to the final version of the manuscript

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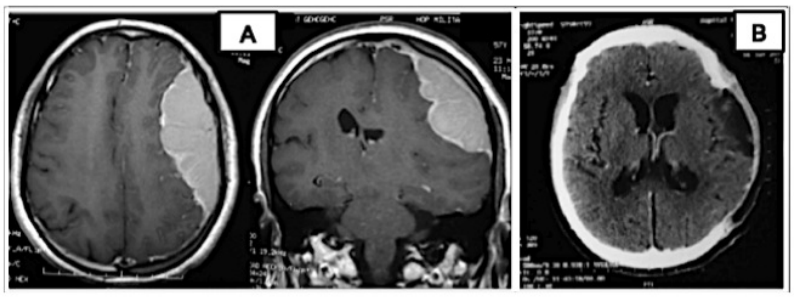

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