Waldenström’s Macroglobulinemia (WM) is an indolent B-cell lymphoproliferative disorder. When there is involvement of the central nervous system, WM is known as Bing Neel Syndrome (BNS). We present a case of BNS, presenting with confusion and left orbital pain and the use of ¹⁸F-FDG PET-CT in making the diagnosis.

This case study demonstrates the ability of ¹⁸F-FDG PET-CT to aid in diagnosing BNS.
Case Report:

A 76-year-old male with known WM, currently in remission, presenting with left eye pain and altered mental status was admitted for further evaluation. Due to concern of possible BNS versus cerebral fungal infection, and contraindication to MRI, FDG PET-CT was performed. The FDG PET-CT (Figures 1A and 1C) revealed two hypermetabolic paraventricular lesions, within the left lateral frontal horn with SUVmax: 10.2 (Fig. 1A); and along the lateral margin of the right lateral ventricle, with SUVmax: 16.1 (Fig. 1C). There was also an FDG avid lesion measuring 1.8 x 0.8 cm with SUVmax: 22.8 (Fig. 1B) in the inferior aspect of the left orbit. These findings are consistent with central nervous system involvement of WM, or BNS (1). Subsequent lumbar puncture revealed atypical lymphocytosis and cytomorphological findings also consistent with involvement of WM (2). The patient subsequently received whole brain and left eye radiation, as well as Rituxumab systemic therapy, with follow up studies demonstrating resolution of the CNS lesions (Figure 2B).

Discussion

BNS is a rare and often a late in disease course (2) manifestation of central nervous system involvement of WM. This syndrome can manifest even during WM remission, and tends to have a worse prognosis compared to those with BNS as the first manifestation of the disease (2,3). Two categories have been described of CNS involvement which are the diffuse form, and the tumoral form, and can present as unifocal or multifocal (as in this case) (3). An appropriate evaluation is necessary to exclude other diseases, such as the hyperviscosity syndrome, which can present with similar neurological deficits, in order for patients to receive optimal treatment (4). Although a gadolinium enhanced MRI should be performed when BNS is suspected (5), in this case it could not be performed due to the prior pacemaker placement.

As demonstrated in this case, FDG PET-CT can be useful in both identifying active disease and for localization of disease, while simultaneously allowing for evaluation of the rest of the body (4).
Conclusion

BNS is a rare and often underdiagnosed disease. FDG PET-CT study can help in the diagnosis and in the precise localization of BNS and therefore it should be considered as an additional diagnostic imaging method to MRI or as an alternative when MRI cannot be performed.
References:


FIGURE 1: Axial CT, PET image, and fused FDG PET/CT of the (A) paraventricular left lateral frontal horn lesion, (B) left orbit lesion, (C) right lateral paraventricular lesion.
FIGURE 2: Contrast enhanced axial CT images. (A) Pre-treatment and (B) post-treatment. The patient underwent ultra-low dose radiation therapy to the whole brain and orbits. He also received 4 cycles of Rituximab. A complete response is seen on the follow up CT (B).