Management of Bing Neel Syndrome

The management of Bing Neel Syndrome (BNS), the disease in which malignant lymphoplasmacytic lymphoma cells invade the central nervous system, has benefitted greatly from the progress made in Waldenström's Macroglobulinemia. In diagnostics, the use of molecular testing for the MYD88 L265P mutation in the spinal fluid proved to be very helpful. For the treatment of BNS, the use of blood brain penetrating drugs like Bruton Tyrosine Kinase (BTK) inhibitors became first choice after the first series of patients treated with Ibrutinib was published in 2019. In the current presentation, the importance of awareness of BNS is stressed, since a delayed diagnosis as well as not establishing a diagnosis in time, can have severe, non-reversible, neurological consequences. In addition, new developments in treatment goals and options will be discussed. With more patient series published treatment results become more clear which can help in managing patients and physicians expectations. Treatment results with the second generation BTK inhibitors Zanubrutinib and Tirabrutinib will be reviewed. Zanubrutinib treatment was given to 21 patients from 5 hospitals. With a median FU of 9.1 months, many data are incomplete but very hopeful was a > 90% clinical response rate at 3 months (EHA poster 2024). Tirtabrutinib was given to 3 patients, and demonstrated clinical and radiological response in all. These BTKi may present new options for BNS patients.