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TREATMENT OF ATYPICAL CNS INFLAMMATION WITH MITOXANTRONE

Khaled Abdel-Aziz, Ian Pomeroy. Walton Centre Foundation Trust

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Case A 42 year old lady with a previous history of Raynaud's syndrome presented with right optic neuritis and right hemiparesis. MRI showed widespread T2 hyperintensities in the deep white matter, corpus callosum, and posterior fossa suggestive of demyelination. LP revealed type 4 oligoclonal bands. Serum pANCA and MPO were transiently elevated. She followed a relapsing remitting course with new right facial numbness, left hemiparesis and new inflammatory lesions seen on MRI imaging over a 4 month period.

Based on a working diagnosis of rapidly evolving severe MS she was treated with three doses of 20mg (12 mg/m²) Mitoxantrone given 4 weeks apart followed my three doses of 10 mg 12 weeks apart. She has been free of new symptoms for 14 month and follow up MRI shown regression of lesions.

Lip biopsy subsequently showed inflammatory infiltrates with a focus score of 3 consistent with a diagnosis of Sjögren's syndrome (SS).

Discussion Treatment of CNS manifestations of SS remains largely empirical. Reported strategies include the use of corticosteroids, cyclophosphamide, methotrexate, azathioprine and cyclosporin. Mitoxantrone represents a possible treatment option in aggressive atypical CNS inflammation where initial diagnostic difficulties are present.