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Pondering Possibilities: PRES vs. PML

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INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) is a rare neuro-radiological syndrome. It can manifest as variable neurological symptoms making it difficult to diagnose.

Although a trigger is usually identifiable, the pathophysiology of PRES remains controversial with uncontrolled hypertension and endothelial dysfunction the main mechanisms implicated.

We describe an unusual presentation of PRES and the associated diagnostic difficulties.

CASE PRESENTATION

A 68 year-old female with a history of Classical Hodgkin's Lymphoma presented with new dizziness, ataxia and progressive diplopia on the background of an active shingles infection.

Treatment comprised of ABVD chemotherapy (doxorubicin, bleomycin, vinblastine, dacarbazine), escalated to PVACEBOP (multi-agent chemotherapy) due to an inadequate interim measure of response by PET. The end of treatment PET was negative, consistent with disease remission.

The following presentation came 3 months after treatment was completed. The patient was lymphopenic with low immunoglobulin's on prophylactic anti-virals (acyclovir and co-trimoxazole).

On presentation she had a markedly ataxic gait, diplopia and cerebellar findings on examination. Her blood pressure was not elevated. She had completed 7 days of oral acyclovir treatment for shingles in the community.

INVESTIGATIONS

A CT head demonstrated no abnormalities. CSF demonstrated a white cell count of 12 with predominance of lymphocytes (Figure 1). CSF protein was slightly elevated at 672. Pending virology, she was commenced on empirical treatment for VZV encephalitis with IV ceftriaxone and IV acyclovir (at a dose of 5mg/kg). Initial virology testing for VZV, HSV1, HSV2 and enterovirus were negative (>95% sensitivity and specificity). With ongoing neurological symptoms an MRI scan was performed (Figure 2). This revealed bilateral extensive white matter changes in keeping with leuco-encephalopathy.

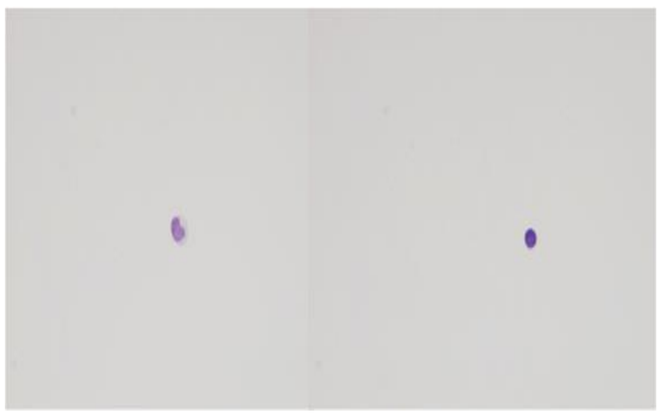

Figure 1: CSF. CSF MGG x 40 high power field.

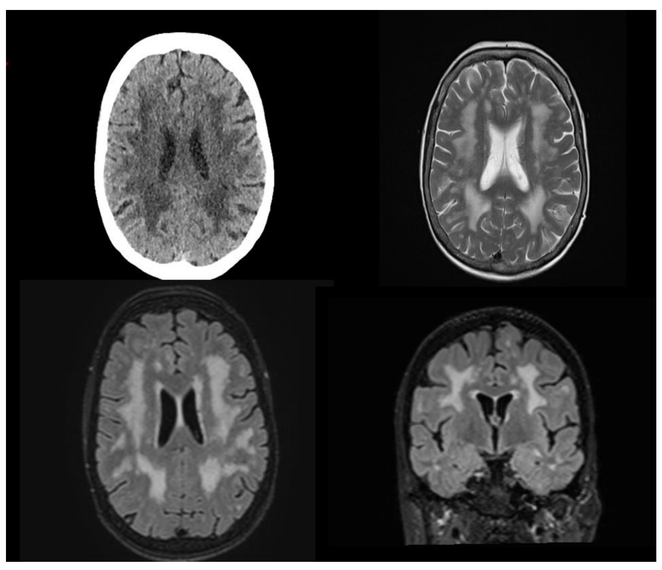

Figure 2: MRI imaging demonstrating bilateral extensive white mater changes

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Differential Diagnosis

In light of the above the working diagnoses included:

- Viral Encephalitis
- PRES
- PML

Treatment and Progression

She remained clinically stable, with subtle neurological improvement demonstrated during her 14 day admission. A further contrast enhanced MRI head reported stable bilateral white matter changes. Extended CSF analysis confirmed the absence of JC Virus, HHV6 and HHV7 infection. A diagnosis of PRES, secondary to chemotherapy was therefore made, supported by her clinical progress. Interval imaging, 6 weeks later, showed stable extensive leukoencephalopathy. The patient made a progressive physical recovery with complete resolution of all clinical signs by 3 months.

DISCUSSION

Neuro-radiological findings are observed in both PRES and PML indicating a clear role for MRI scanning. MRI in patients with PRES commonly demonstrates bilateral, symmetric cerebral white matter oedema with subcortical and cortical involvement. PRES frequently involves the posterior cerebral hemispheres and the parietal and occipital lobes in 70% of cases. In PML the MRI is characterized by subcortical scalloped appearances over the white matter region. These are not conformed to a single vascular territory, and are associated with minimal enhancement/mass effect[1].

Chemotherapy was considered the precipitating cause of PRES in our patient. In 2018, Mahmoud, Ahmed and Rand outlined a case of PRES occurring in a patient with Hodgkin's Lymphoma after being treated with Adriamycin, Bleomycin, Vinblastine and Dacarbazine Chemotherapy[2]. Our patient completed chemotherapy 3 months prior to presentation, in retrospect she reported mild visual symptoms preceding her acute presentation by several months. This subacute onset of symptoms supported PRES rather than PML as a final diagnosis.

CONCLUSIONS

This case highlights the importance of considering rare neurological causes in immunosuppressed patients presenting with new neurological symptoms. It also highlights the benefits of MRI in the assessment of patients with new neurological symptoms.

Both PRES and PML are now linked with new chemotherapy and immunotherapy regimens; thus clinicians involved in the use of such medicines should be aware of their potential significant adverse effects, and remain vigilant when such complications arise.

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