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CONTROVERSIES IN MULTIPLE MYELOMA (COMy)

A CASE OF MYELOMATOUS MENINGITIS

A rare clinical entity!

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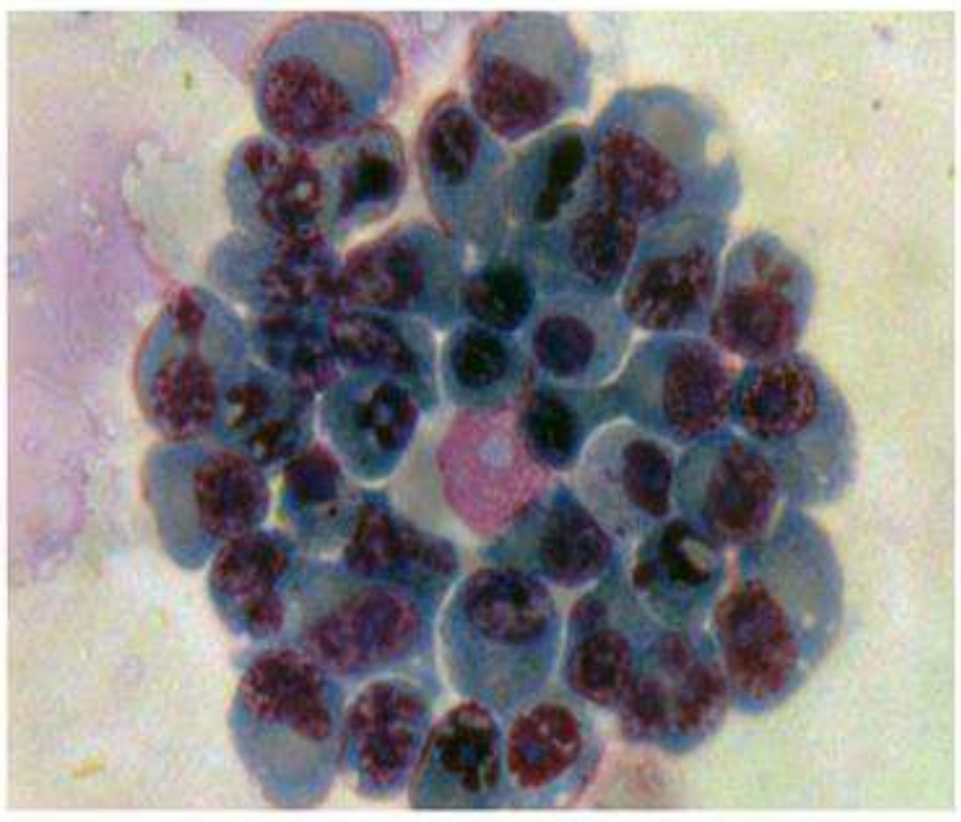
Introduction

A 48-year old man with multiple myeloma (MM) who was treated with various forms of targeted therapy, chemotherapy and haematopoietic stem cell transplantations (SCT) presented to hospital with short episodes of pins and needles in his left hand. It was associated with episodes of neck spasm, severe headache, nausea and aphasia which lasted 10-15 minutes. His MRI scan was unremarkable but he developed generalised tonic-clonic seizures, nystagmus, aphasia and loss of consciousness after one week.

Results

His CT brain at this stage was unremarkable and he was noticed to have aphasia with agitation. A lumbar puncture was performed and cerebrospinal fluid (CSF) fluid was clear but his biochemistry showed a blood cell count (WBC) of 250 cells/uL (normal range: 0-5 cells/uL), with 99% neutrophils (normal value: <2 polymorphonucleocytes), red blood cell count (RBC) <1 cell/uL and absence of bacteria; no growth after 72-hour incubation. Meningococcal and pneumococcal PCR tests were both negative.

His CSF cytology revealed clusters of abnormal plasma cells with multinuclear forms without red cell contamination, a picture consistent with myelomatous meningitis [Figure1]. Flow cytometry analysis confirmed these findings as it revealed that 90% of the CSF white cells had the following immunophenotype CD19- CD38+ CD138+ CD117+ [weak] CD56+.



Cerebrospinal Fluid (CSF) cytology post-lumbar puncture revealing the presence of atypical plasma cells with blast features (x100).

Conclusion

A diagnosis of myelomatous meningitis was concluded. He was treated symptomatically with analgesics, benzodiazepines and levetiracetam to control his seizures. As a result of the poor prognosis agreed upon by the multidisciplinary team (MDT), no trial treatment, such as intrathecal chemotherapy or radiotherapy was commenced.

Discussion

Myelomatous meningitis is clearly a rare complication of multiple myeloma that can be challenging to recognise and definitively diagnose. In clinical practice the suspicion for CNS infiltration is very low as the incidence of CNS involvement is approximately 1%. In the presence of progressive neurological deficits in patients with MM, myelomatous meningitis should be suspected and MRI scan considered promptly. CSF cytology is essential to establish a definitive diagnosis and should be carried out even in cases where the brain MRI is reported as normal. There is currently no definitive treatment for myelomatous meningitis.

Clinicians should be aware of the unspecific neurological deficits that could form the presenting symptoms of the disease so that appropriate investigations, including brain MRI and CSF cytology, are carried out promptly to establish a swift diagnosis.

Unfortunately, the pathophysiology of this complication is still poorly understood and there is no possible curative treatment. In a few cases intrathecal chemotherapy was used as therapy but showed poor results. Myelomatous meningitis has an extremely poor prognosis, even after intrathecal chemotherapy or craniospinal radiation. Overall survival and prognosis remained poor highlighting a need for further data from future research studies with a view to improve clinical outcomes.

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