European Journal of Case Reports in Internal Medicine

Uveo-Meningitis and Myelodysplastic Syndrome

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Doi: 10.12890/2022_003633 - European Journal of Case Reports in Internal Medicine - © EFIM 2022

Received: 09/10/2022
Accepted: 12/10/2022
Published: 31/10/2022

How to cite this article: Ferrão D, Ribeirinho-Soares P, Almeida J. Uveo-meningitis and myelodysplastic syndrome. *EJCRIM* 2022;**9**: doi:10.12890/2022_003633.

Conflicts of Interests: The authors declare there are no competing interests.
Patient consent: Obtained.

ABSTRACT

Uveo-meningeal syndromes are any disorder characterized by the involvement of the uvea and the meninges. They can have multiple causes, including infectious, autoimmune and malignant diseases. We report the case of a patient with a 10-year-old diagnosis of myelodysplastic syndrome that had been stable. He presented with new onset uveitis, ataxia, diplopia and fluctuating consciousness level, without any obvious cause revealed by brain imaging or blood chemistries. An extensive aetiological search showed no obvious cause. Initially, the patient improved spontaneously and was discharged. However, 2 months later, he deteriorated once again, this time with disperse adenomegalies that were not present previously. The bone marrow biopsy showed a high number of blasts, which affirmed the progression of the previously known myelodysplastic syndrome. The lymph nodes were not biopsied due to very low life expectancy, making the procedure futile. A presumptive diagnosis of uveo-meningitis caused by paraneoplastic syndrome was made. The patient died a few months later.

LEARNING POINTS

- Uveo-meningeal syndrome (UMS) is a rare entity usually of infectious or inflammatory aetiology.
- UMS may be caused by some neoplastic disorders, mainly lymphoma and metastasis of solid tumours; myelodysplastic syndrome (MDS) has not been previously implicated.
- Diagnosis should include lumbar puncture and a thorough search for the aetiology; treatment is directed towards the aetiology.

KEYWORDS

Uveo-meningeal syndrome, myelodysplastic syndrome

INTRODUCTION

The uveo-meningeal syndromes (UMS) are a heterogeneous group of disorders that share the feature of concomitant involvement of the uvea and the meninges [1]. Several diseases can cause UMS, including infectious, autoimmune and neoplastic disorders, some of which are not obvious or have a remission-relapsing pattern, posing a diagnostic challenge. We report the rare case of a paraneoplastic uveo-meningeal syndrome caused by myelodysplastic syndrome (MDS).

CASE DESCRIPTION

We describe an 83-year-old man with a history of hypertension, dyslipidaemia and MDS diagnosed 10 years previously, with no cytogenetic changes and that had been stable with no treatment. He was taken to our hospital due to persistent headaches and new onset ataxia and diplopia. The patient, previously independent in daily activities, now needed a wheelchair and help for most activities. On admission, he had ataxia and VI nerve palsy. Ophthalmological evaluation showed the presence of uveitis. A cerebral computed tomography (CT) scan

and magnetic resonance imaging (MRI) were performed and did not show any changes. A lumbar puncture (LP) was also performed, showing an inflammatory cerebrospinal fluid (CSF) with 100 white cells (99% lymphocytes), a protein level of 1.34 g/l (cut-off 0.45 g/l) and normal glucose. At this point, we assumed the presence of a UMS of unknown aetiology. Blood cultures were negative. CSF was further analysed, and bacteriological, virological and mycological tests were negative. CSF immunocytochemistry was not suggestive of central nervous system involvement by the MDS. A complete immunological panel was negative. A thoraco-abdominopelvic CT scan showed several retroperitoneal lymphadenopathies, the largest of which measured 18×13 mm, without any lung or abdominal changes suggestive of sarcoidosis or tuberculosis. The search for mycobacterium in the CSF, blood and sputum was also negative. The complete blood count (CBC), the peripheral blood smear (PBS) and cell immunophenotyping were normal. A lymph node biopsy was not performed due to spontaneous improvement of the patient. He maintained a slightly unsteady gait but without diplopia or headaches. Therefore, he was discharged with the diagnosis of UMS of unknown cause with spontaneous improvement. However, 2 months later, he had a relapse of the headaches and a new-onset confusional state. All the above-mentioned studies were repeated, including the CT scan, MRI and CSF analysis, and findings were all identical to the first results. The thoraco-abdominopelvic CT scan showed an increase in the size of the retroperitoneal lymphadenopathies and de novo cervical lymphadenopathies, the largest of which measured 19×9 mm. The CBC and PBS remained normal but now the possibility of progression of the MDS was greater. A medullary biopsy was performed, showing 16% blast cells. It was concluded that the MDS was progressing to high-risk disease, which meant an average life expectancy of 9 months. Due to this fact and the advanced age and frailty of the patient, it was decided to pursue a conservative approach and the lymph node biopsy was deferred. The patient was discharged and died a few weeks later.

DISCUSSION

The association of uveitis with meningoencephalitis is uncommon and warrants a thorough search for an underlying aetiology. Most of the time, the anatomical continuity between the meninges and the choroid is the main reason for the association of the two conditions. In rarer cases, meningitis and uveitis are associated through the systemic dissemination of a common pathological process [2]. Most UMS are of an infectious nature. Potential bacterial causes include *Bartonella*, tuberculosis, syphilis, Lyme disease and Whipple disease. Fungi such as *Candida albicans* and *Aspergillus* are also potential causes, as are viruses like herpes simplex, varicella-zoster and cytomegalovirus [3]. In our case, all CSF, blood cultures and serologies were negative. Furthermore, the patient had some clinical characteristics that made some of these aetiologies, such as Lyme disease, less likely, and as he was not immunocompromised, we did not need to consider fungal aetiologies. Tuberculosis was a possibility, but the search for *Mycobacterium tuberculosis* was persistently negative. In terms of possible inflammatory causes, the immunological panel was negative, including for IgG4-related disease, sarcoidosis, Behçet syndrome and granulomatosis with polyangiitis, and he lacked other systemic manifestations of these diseases. Vogt-Koyanagi-Harada syndrome (VKHS) was another possibility; however, it is usually accompanied by cutaneous and inner ear complaints, which our patient did not have. Furthermore, this diagnosis is one of exclusion, requiring that all other causes of UMS be ruled out [4]. Neoplastic disorders are among the less common causes of UMS and include haematological tumours, such as lymphoma, as well as solid tumours, most commonly metastasis to the eye.

Several case reports of UMS have been published in the literature, most of which had an infectious aetiology. A review of 110 patients [2] showed that most cases of UMS were caused by VKHS, syphilis or sarcoidosis, or were idiopathic. There are also some published cases with neoplastic aetiologies, mainly lymphoma and metastatic carcinoma. Scarce reports mention paraneoplastic syndromes, usually in small-lung cell carcinoma or thymoma. To our knowledge, no cases of UMS caused by MDS have been reported.

The first issue worth discussing is the rarity of UMS with a neoplastic origin, especially caused by MDS. In our case, there were no peripheral signs of active MDS and medullary and ganglia studies were not pursued due to spontaneous improvement of the patient. Two months later, the high percentage of bone marrow blasts confirmed the diagnosis of high-risk MDS. This was, very likely, the aetiology behind the UMS. A second important issue is the diagnostic algorithm in these patients. While in patients with neurological manifestations, LP seems unavoidable, in patients with plain uveitis it does not seem to change the outcome or may even lead to excessive futile testing [2, 5]. After the diagnosis of UMS, the definitive aetiological diagnosis must always be chased, since the treatment is tailored to it. In our patient, treatment was not initiated due to his fragility and the poor prognosis of the disease. Lastly, clinical fluctuation in these patients is not usual, and the fact that it happened in our case may have masked the real aetiology behind the UMS.

In conclusion, this case emphasizes the importance of always considering unusual presentations of certain illnesses. MDS is a common diagnosis in the elderly, can progress even when stable for many years and can present in very unusual ways.

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Internal Medicine

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