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Aseptic meningitis and hydrocephalus secondary to neurosarcoidosis

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SUMMARY

A 53-year-old woman presented to hospital with gait instability, urinary incontinence and confusion. She had a 4-month history of headache, blurred vision, personality change and memory problems. Magnetic Resonance Imaging of the brain after contrast application showed tectal plate and occipital enhancement, as well as a known hydrocephalus. Cerebrospinal fluid showed aseptic meningitis with no evidence of clonal expansion. After further imaging that showed generalised lymphadenopathy and subsequent tissue biopsy that showed granulomatous lymphadenitis, she was diagnosed with neurosarcoidosis. She was treated with steroids which resulted in immediate cognitive and motor improvements as well as resolution of her urinary incontinence. We discuss the features of this case that pointed towards neoplastic, infective and other autoimmune aetiologies. We describe how they were excluded and provide the rationale for our treatment. This case demonstrates an important sequela sarcoidosis, and we conclude by recommending a multidisciplinary approach towards its diagnosis and management.

BACKGROUND

Sarcoidosis is a multisystemic granulomatous disorder. Its annual incidence in the United Kingdom is between 9.7 and 14.5 per 100 000.^{1 2} Diagnosis of this condition requires the exclusion of other granulomatous disorders such as lymphoproliferative disorders, infections, drug reactions and other autoimmune conditions. Neurological involvement (neurosarcoidosis) occurs in approximately 5%–26% of cases^{3 4} and may result in peripheral or central nervous system (CNS) manifestations.

CASE PRESENTATION

In September 2020, a 53-year-old Caucasian woman was admitted to hospital with gait instability, urinary incontinence and confusion. She had a history of hypertension for which she was taking antihypertensives and a maternal history of breast cancer which was diagnosed in the seventh decade. Her symptoms started in the preceding May with a sudden headache and associated blurred vision, which was treated as a migraine. However, over the next 3 months, her headache persisted. During this time, she also developed memory problems. A Magnetic Resonance Imaging (MRI) scan of the brain in August revealed dilated ventricles consistent with hydrocephalus (figure 1). When she was admitted to hospital, on examination, she had mild gait instability and her blood pressure was 220/142 mm Hg. Her routine blood tests were

normal aside from a mild lymphopaenia which was present from the onset of her symptoms in May. Serum Angiotensin Converting Enzyme (ACE) was non-elevated at <12 units/L (normal range (NR): 20–70). Her Addenbrooke's Cognitive Examination-Revised (ACE-R) score was 45/100. Her cerebrospinal fluid (CSF) showed an elevated white cell count of 60 cells/ μ L (NR: 0–5), an elevated protein of 1.04 g/L (normal range: 0.15–0.45) and a low glucose of 1.6 mmol/L (NR: 2.2–4.0). Subsequent CSF flow cytometry confirmed lymphocytosis with T-cell predominance with no phenotypic evidence of an atypical lymphoid infiltrate. Whole-body imaging showed widespread lymphadenopathy (figures 2 and 3). Imaging of the brain showed enhancement in the occipital lobe and tectal plate (figure 4). A subsequent core biopsy of the right inguinal lymph node showed granulomatous lymphadenitis (figure 5). She underwent extensive testing for infective pathogens and an autoimmune screen, both of which were negative.

DIFFERENTIAL DIAGNOSES

Her differential diagnoses are discussed in table 1.

The successful exclusion of the aforementioned diagnoses, along with the findings of her lymph node biopsy and focal areas of cerebral enhancement,

Figure 1 Axial T2 MRI of the brain without contrast shows ventricular enlargement consistent with hydrocephalus (August 2020).

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Figure 2 Images (A) and (B) show bilateral hilar lymphadenopathy on chest X-ray and computerised tomography (CT) of the chest, respectively (October 2020).

allowed us to arrive at the diagnosis of neurosarcoidosis. The lack of a confirmatory test in diagnosing sarcoidosis means that it remains a diagnosis of exclusion. It is notoriously difficult to ensure that tuberculosis has been excluded, with lymphoma being the other major diagnosis to rule out. The advent of CSF flow cytometry has been very helpful in the latter. Excluding lymphoma was particularly important here as there is a two-way statistical association between an individual and a first-degree relative for breast cancer and non-Hodgkin's lymphoma.⁵ The Neurosarcoidosis Consortium Consensus Group published diagnostic criteria in 2018 for possible, probable and definite neurosarcoidosis.⁶ As per those criteria, the index case qualifies for a diagnosis of probable neurosarcoidosis. To establish a diagnosis of definite neurosarcoidosis, a CNS biopsy would have been required. However, this is an invasive procedure that carries risk. As we had rigorously excluded other diagnoses, we felt confident that the diagnosis we had established was accurate.

Figure 3 CT of the abdomen shows right inguinal lymphadenopathy which was biopsied (October 2020).

Figure 4 Images (A) and (B) show tectal plate and occipital enhancement on sagittal and axial T1 postcontrast MRI of the brain, respectively (October 2020).

Hence, we did not feel that subjecting this patient to an invasive CNS biopsy was justified at this time.

TREATMENT

The patient was treated with 3 days of 1 mg methylprednisolone intravenously, followed by 60 mg of daily oral prednisolone. After 5 days of steroid treatment, her ACE-R score increased to 75/100. Her mobility also improved such that it was possible

Figure 5 The lymph node core biopsy demonstrates effacement of the nodal architecture by closely packed non-caseating granulomas seen mainly in the upper core. The lower core shows zones of associated hyalinising fibrosis (star symbol). Numerous Langhans-type giant cells are seen (insert, October 2020).

Table 1 Evaluation and exclusion of differential diagnoses

Differential diagnosis	Features in favour	How diagnosis was excluded
Lymphoma	<ul style="list-style-type: none"> CSF findings of lymphocytosis with raised protein Widespread lymphadenopathy on CT of the chest, abdomen and pelvis Peripheral lymphopaenia since the onset of symptoms ranging between $0.5 \times 10^9/L$ and $1.0 \times 10^9/L$ (NR: $1.4\text{--}4.8 \times 10^9/L$) Maternal history of breast cancer 	<ul style="list-style-type: none"> Absence of atypical cells on CSF flow cytometry No evidence of lymphoma on tissue biopsy
Chronic pathogenic infections	<ul style="list-style-type: none"> Subacute history with neurological symptoms Lymph node biopsy showing granulomatous lymphadenitis CSF showing raised lymphocytes, low glucose and raised protein suggestive of tuberculosis or fungal infection 	<ul style="list-style-type: none"> Negative CSF, serum, urine and sputum analysis for pathogens, including acid-fast bacillus stains, prolonged culture and mycobacterium tuberculosis PCR on CSF Negative staining for microorganisms on lymph node biopsy
Carcinomatous meningitis	<ul style="list-style-type: none"> Subacute history with neurological symptoms and aseptic meningitis 	<ul style="list-style-type: none"> Absence of malignancy identified on CT of the chest, abdomen and pelvis/ MRI of the brain and spine No malignant cells in CSF
Autoimmune vasculitis	<ul style="list-style-type: none"> Lymph node biopsy showing granulomatous lymphadenitis 	<ul style="list-style-type: none"> Negative auto-antibody screen
Phaeochromocytoma	<ul style="list-style-type: none"> Headache and visual disturbances Labile blood pressures ranging between 191/114 mm Hg and 220/142 mm Hg 	<ul style="list-style-type: none"> Normal 24-hour urinary metanephrines Absence of adrenal mass on CT of the chest, abdomen and pelvis

CSF, cerebrospinal fluid; NR, normal range.

to discharge her home. At the time of discharge, the patient's cognitive problems had significantly improved although she continued to experience mild gait instability. She was discharged on prednisolone 60 mg daily, and this was reduced to 40 mg daily over the next 4 weeks. Four weeks after discharge, she was also started on azathioprine 25 mg daily with a plan to increase to 150 mg daily.

In the context of sarcoid-related hydrocephalus, we could not find any published data as to whether medical or surgical management or a combination of the two, would be the most appropriate treatment. In her case, the rapid response to steroids meant there was no indication for acute neurosurgical intervention. Moreover, active inflammation can also block

ventriculo-peritoneal (VP) shunts, meaning that the decision to offer early neurosurgical intervention should be taken carefully. Nonetheless, on discharge, this patient was placed under the care of a specialist multidisciplinary team that did include a neurosurgical opinion. The plan was to consider neurosurgical intervention if medical management failed to control her hydrocephalus-related symptoms. Although hydrocephalus is reported to be present in only 6% of cases of neurosarcoidosis,⁴ it is becoming increasingly recognised as a feature of this condition. From our literature review, we found 21 cases of hydrocephalus identified as a presenting feature of neurosarcoidosis. Of these 21 patients, 17 underwent both medical and surgical management for their hydrocephalus, 2 underwent surgery alone

Table 2 Summary of treatment of published cases of neurosarcoidosis with hydrocephalus (adapted from Saban *et al*²⁷)

Author	Treatment	Outcome
Pandey <i>et al</i> (2021, index case)	Corticosteroids + azathioprine with provisional plan for surgical intervention	Partial recovery
Saban <i>et al</i> ²⁷ (2020)	Corticosteroids + methotrexate, followed by VP shunt months later	Partial recovery
McKeever <i>et al</i> ¹¹ (2019)	Case 1: Corticosteroids + azathioprine Case 2: Initial endoscopic third ventriculostomy followed by multiple neurosurgical procedures 7–10 years later, including a VP shunt insertion, two shunt revisions and endoscopic fenestration of the third and fourth ventricles	Case 1: Complete recovery Case 2: Partial recovery after the initial procedure but significant neurological disabilities after 10 years
Sugiyama <i>et al</i> ²⁸ (2016)	Corticosteroids + VP shunt	Partial recovery
Chandna <i>et al</i> ²⁹ (2015)	Corticosteroids + VP shunt	Death
Hitti <i>et al</i> ³⁰ (2015)	Corticosteroids + VP shunt + mycophenolate mofetil	Unknown
Matsuda <i>et al</i> ³¹ (2015)	Corticosteroids + ventriculostomy followed by VP shunt	Complete recovery
Sano <i>et al</i> ³² (2015)	Corticosteroids + VP shunt + methotrexate + infliximab	Partial recovery
Yoshitomi <i>et al</i> ³³ (2015)	Corticosteroids + endoscopic fenestration foramen of Magendie, followed by VP shunt	Complete recovery
Tabuchi <i>et al</i> ³⁴ (2013)	Corticosteroids + VP shunt	Partial recovery
Zoja <i>et al</i> ⁷ (2012)	Not applicable as diagnosis established at autopsy	Death
Kim <i>et al</i> ³⁵ (2012)	Corticosteroids + VP shunt	Complete recovery
van Rooijen <i>et al</i> ⁸ (2011)	VP shunt + corticosteroids	Partial recovery
Berhouma <i>et al</i> ³⁶ (2009)	Corticosteroids + right temporal tip lobectomy	Complete recovery
Brouwer <i>et al</i> ³⁷ (2009)	Ventriculocopy-assisted fenestration of lateral ventricle cyst	Complete recovery
Westhout <i>et al</i> ³⁸ (2008)	Corticosteroids + VP shunt	Complete recovery
Benzagmout <i>et al</i> ²⁰ (2007)	Corticosteroids + external ventricular drain	Partial recovery
Muayqil <i>et al</i> ³⁹ (2006)	Corticosteroids + VP shunt	Partial recovery
Muniesa <i>et al</i> ⁴⁰ (2006)	Corticosteroids + VP shunt	Complete recovery
Onoda <i>et al</i> ⁴¹ (2004)	Corticosteroids + VP shunt	Death from nosocomial pneumonia
Chiang <i>et al</i> ⁴² (2002)	Corticosteroids + VP shunt	Unknown

VP, ventriculo-peritoneal.

Case report

Table 3 Sequential ACE-R assessment scores.

ACE-R domain	October 2020	November 2020	January 2021
Attention (/18)	4	14	13
Memory (/26)	11	18	18
Fluency (/14)	1	5	2
Language (/26)	20	24	26
Visuospatial (/16)	9	14	16
Total (/100)	45	75	76

ACE-R, Addenbrooke's Cognitive Examination-Revised.

and 1 underwent medical management alone. The remaining patient died suddenly and did not receive any treatment as her hydrocephalus was established at autopsy.⁷ These results are summarised in [table 2](#) along with the index case. Despite the lack of data on the management of sarcoid-related hydrocephalus, most authors employed the use of medical management prior to surgical management, as in this case. One author even suggested that earlier use of steroids may have precluded the need for surgical placement of a VP shunt altogether.⁸

In general, the management of sarcoidosis can vary depending on the organ system involved. For example, the British Thoracic Society recommends that patients with pulmonary sarcoidosis can be managed without treatment if they remain asymptomatic due to high rates of spontaneous remission.⁹ However, neurosarcoidosis rarely undergoes spontaneous remission and remains a severe illness, often requiring long-term treatment. A step-wise approach to management includes using steroids as initial management. The next step up includes methotrexate, mycophenolate mofetil, leflunomide and azathioprine, before finally escalating to biological agents such as infliximab, adalimumab and rituximab.¹⁰

OUTCOME AND FOLLOW-UP

The patient was reviewed 7 weeks after being discharged. At this time, her ACE-R score was 76/100. Her sequential ACE-R assessments with breakdown of scores are shown in [table 3](#).

DISCUSSION

It is important to reiterate that establishing a diagnosis of neurosarcoidosis can be challenging and time-consuming. We identified one case where a diagnosis of neurosarcoidosis was established over 10 years after the identification of hydrocephalus. Unfortunately, that patient developed progressive disabilities that did not respond to initial therapy. The authors concluded that establishing a swifter diagnosis could have prevented irreversible neuronal damage, thus highlighting the importance of a timely diagnosis.¹¹ Histologically, sarcoidosis is characterised by the presence of non-caseating granulomas. A number of immune cell types are found in these granulomas, including macrophages, epithelioid cells and multinucleated giant cells, as well as lymphocytes that are the predominant cell type in the central part of a sarcoid granuloma.¹² Accumulation of activated T cells to the sites of inflammation causes a peripheral lymphopaenia which is seen in the majority of patients with sarcoidosis including this case.¹³⁻¹⁵ The epithelioid cells secrete ACE, and this enzyme is widely used as a biomarker in the work-up for sarcoidosis. However, its use is limited, as quoted sensitivity and specificity for elevated ACE in diagnosing sarcoidosis are 41.4% and 89.9%, respectively.¹⁶ Another series of 128 patients with neurosarcoidosis found that CSF protein was raised in 76% of samples, as in this case, with a median CSF protein level of 0.8 g/L, though it may be significantly elevated.¹⁷

Finally, some of the more common manifestations of neurosarcoidosis include cranial neuropathy, peripheral neuropathy, mononeuropathy, myopathy, psychiatric disorders and cerebellar ataxia.⁴ The pathophysiological mechanisms leading to these manifestations are not fully understood, although upregulation of inflammatory cytokines such as tissue necrosis factor α , oxidative damage and alterations in neurotransmitter metabolism are thought to contribute to cognitive deficits.^{18 19} Inflammation of the arachnoid villi may lead to reduced CSF absorption, causing a communicating hydrocephalus and its associated clinical features.^{20 21} The patient's elevated blood pressure (ranging between 191/114 mm Hg and 220/142 mm Hg) was felt to be due to her poor compliance with antihypertensives secondary

Patient's perspective

My husband and I started to notice over a year ago that my movement was becoming languid and whilst in [censored location] for my son's graduation, I tripped on a pavement outside our hotel and could not prevent my head from colliding with the pavement, which caused an injury to the bridge of my nose. In the months after this, my movement became worse. My husband and I are [censored sporting event] ticket holders; it became very uncomfortable to walk from the car park to the stadium. I visited the doctor on a few occasions as my joints, particularly knees, shoulders and ankles, became extremely painful. I was almost constantly also suffering from a severe headache. Various tests could not find any problems to explain these pains. During early May, I woke with blurred vision and an extremely bad headache; we were told to go to [censored location] where they would carry out a CT scan. My memory was getting worse at this point and my employer was raising concerns. My general practitioner (GP) arranged for a mental assessment, more blood tests and an MRI scan. We met with the neurologist, and they suggested that the problems were due to migraines. My employer decided that I was not able to continue to work and wrote to my GP at the end of August expressing concern that there was something seriously wrong which needed to be looked into.

In September, I became more confused and found that I was not able to do basic things like write greetings cards or emails; my husband thought my driving was less controlled and I was very light headed and in a lot of pain in my joints. I fell over in our bedroom and was too feeble to be able to get to my feet. About a week after this, I went for a hair appointment and got confused and very light headed whilst in the shopping centre. My husband and my employer all realised that something was very wrong and in late September contacted my GP again. My GP said that [(s)he] wanted me to be admitted to hospital, so my husband took me in. I spent almost 9 weeks in hospital; my memory became much worse and I was telling my husband that I was thinking I was in a spa on some days and often mentioned that I had been talking to my late mum and dad. I contracted COVID-19 in hospital. Although I did fall over a few times whilst in hospital, and on a couple of occasions was left in a very uncomfortable position for a considerable time, I have to say that the care I received was absolutely brilliant and I want to thank all involved for getting to the bottom of my illness and diagnosing my condition. Everybody at [censored location] was so caring and professional despite these very difficult times. I am at home now; I am still having problems with my mobility, but my memory and confusion are vastly improving.

Learning points

- This case demonstrates an important sequela of sarcoidosis.
- The lack of a confirmatory test and the multisystemic nature of sarcoidosis can make its diagnosis very challenging.
- There is a lack of evidence as to whether medical or surgical management or a combination of the two, would be the most appropriate treatment in sarcoid-related hydrocephalus.
- Prompt diagnosis may result in more favourable outcomes, and a multidisciplinary approach towards diagnosis and management is recommended.

to her cognitive impairment. Hypertension can also occur in sarcoidosis secondary to autonomic dysfunction caused by a small fibre neuropathy^{22–25} or due to renal dysfunction caused by interstitial granulomatous nephritis or other glomerular pathologies.²⁶ There was no evidence of renal involvement in this patient at the time this report was written.

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