Sarcoidosis presenting with acute hydrocephalus in a New Zealand European female
Arjun Chandna, Christopher Todd, David Murphy, Ronald Boet, Roderick Duncan

Abstract

Hydrocephalus as the presenting feature of sarcoidosis is extremely rare. We report the case of a 41-year-old New Zealand European female who presented with acute hydrocephalus as the first manifestation of systemic sarcoidosis.

Sarcoidosis is a multisystem granulomatous disorder of unknown aetiology. Neurosarcoidosis is reported in only 5–10% of cases. If neurological involvement occurs, it tends to happen early in the natural history of the disease: diagnosis can be challenging and neurosarcoidosis is frequently under-recognised.

We present a case of acute hydrocephalus as the first manifestation of systemic sarcoidosis. To our knowledge this is the first such case described in New Zealand.

Case report

A 41-year-old New Zealand European female was admitted to hospital after the incidental finding of a blurred right optic disc margin during routine diabetic retinal screening (Figure 1). CT brain scan demonstrated “acute hydrocephalus and multiple enhancing lesions consistent with intracerebral and leptomeningeal metastases”.

Figure 1. Right retinal photograph. Blurred right optic disc margin detected during routine retinal photography performed as part of diabetic screening service
On admission to neurosurgery she reported a 2-month history of lethargy, anorexia and intermittent early morning vomiting. More recently she had become unsteady on her feet, developed diplopia and right-sided weakness. Her past medical history included diabetes mellitus type 2, hypertension and asthma.

On examination she had mild weakness in her right leg, with spasticity, sustained clonus and upgoing plantar reflex. She reported diplopia in all directions of gaze.

Gadolinium-enhanced MRI scan of her brain and spine showed “acute hydrocephalus with diffuse nodular enhancing lesions throughout the neuraxis including at the cerebral aqueduct” (Figure 2). Differential diagnoses included carcinomatous meningitis, tuberculosis and sarcoidosis.

Figure 2. Gadolinium-enhanced post-contrast T1-weighted MRI scan of brain and thoracic spine. Sagittal sections of T1-weighted MRI scan demonstrating acute hydrocephalus and diffuse nodular enhancing lesions throughout the neuraxis, including at the cerebral aqueduct and foraminae of Luschka and Magendie

A chest radiograph and subsequent CT scan confirmed bilateral hilar and mediastinal lymphadenopathy. Serum calcium was 2.5mmol/L (range 2.2–2.6), angiotensin converting enzyme 30IU/L (range 12–52). Quantiferon-TB® test, and tumour marker panel were negative.

She was commenced on 8mg oral dexamethasone and referred to oncology to investigate a possible malignant cause, where mediastinal lymph node biopsy confirmed granulomatous inflammation consistent with sarcoidosis (Figure 3).

With a diagnosis now ascertained, neurology were consulted and advised 1g intravenous methylprednisolone followed by 60mg oral prednisone. During the following 8 days the patient deteriorated with imaging confirming progression of hydrocephalus. An external ventricular drain was inserted and later replaced with ventriculoperitoneal shunting.

Subsequent improvement in her systemic and neurological symptoms was accompanied by interval reduction in ventricular size and the patient was discharged.
Two days post-discharge she represented with tachypnoea. A CT pulmonary-angiogram confirmed extensive bilateral pulmonary emboli with features of right heart strain. Sadly, resuscitation attempts, including intravenous thrombolysis, were unsuccessful and the patient died.

Figure 3: Haematoxylin and eosin stained section of mediastinal lymph node. Histology from mediastinal lymph node demonstrating non-caseating granulomatous inflammation consistent with sarcoidosis

Discussion

Neurosarcoidosis is often considered in the differential diagnosis for patients who present with neurological symptoms in the context of a known history of sarcoidosis. Unfortunately however, neurosarcoidosis rarely has a systemic harbinger, rendering diagnosis difficult. This case highlights the importance of obtaining a histological diagnosis in patients with suspected neoplastic disease.

Involvement of the nervous system is reported in 5–10% of cases of sarcoidosis, however post-mortem studies suggest this is an underestimate. A recent Australian study reported neurological involvement in 26% of patients with sarcoidosis.

Hydrocephalus as the first manifestation of sarcoidosis is rare: a review of the literature reveals six previous documented cases. Five of these patients were male and none were Caucasian (2 African-American, 2 Korean, 1 Japanese and 1 Malian). The only case of neurosarcoidosis described in the New Zealand medical literature is that of a 48-year-old woman presenting with a vagal mononeuropathy.
Sarcoidosis carries significant morbidity. Patients with neurological manifestations, particularly hydrocephalus, have increased mortality and are more likely to relapse on treatment. However new treatments are emerging and hence early diagnosis is crucial.

Criteria are available to assist with the diagnosis of neurosarcoidosis but a high index of suspicion is required in patients with otherwise unexplained neurological symptoms. Increased awareness amongst clinicians may lead to earlier diagnosis and treatment.

**Author information:** Arjun Chandna1*, Christopher Todd1; David Murphy1; Ronald Boet2; Roderick Duncan1.

1) Department of Neurology, Christchurch Public Hospital, Christchurch, New Zealand
2) Department of Neurosurgery, Christchurch Public Hospital, Christchurch, New Zealand

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**Correspondence:** Dr Arjun Chandna. Email: arjunchandna@gmail.com

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