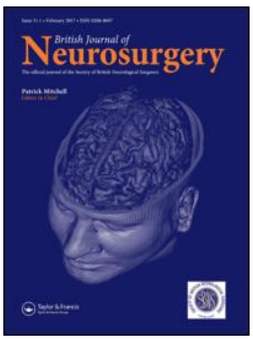

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SHORT REPORT

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A case of primary central nervous system lymphoma presenting as a shunt complication

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ABSTRACT

The authors describe an 82-year-old female with a right frontal ventriculoperitoneal (VP) shunt for long-standing normal pressure hydrocephalus (NPH) who presented with worsening incontinence and gait instability. She was found to have right lateral ventricle collapse around the shunt catheter and subsequently underwent shunt revision, which failed to improve her symptoms. Magnetic resonance imaging (MRI) was obtained on postoperative day two, which demonstrated a ventricular lesion. Endoscopic brain biopsy was performed and a diagnosis of primary central nervous system lymphoma (PCNSL) was made. The authors believe this is the first published case of PCNSL presenting as a VP shunt complication in a patient with NPH.

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Brain tumor; lymphoma; oncology; primary CNS lymphoma; intraventricular

Introduction

Primary central nervous system lymphoma (PCNSL) accounts for 3–5% of primary brain tumors, with an annual incidence of five per one million person-years.[1–3] It is a category of extranodal non-Hodgkin lymphoma located within the brain, eyes, leptomeninges, and spinal cord.[1–3] In 95% of cases, PCNSL consists of diffuse, large B-cell infiltrates. The remaining cases are either low-grade B-cell or T-cell origin.[1,3] PCNSL is usually associated with immunosuppression but has recently been found with increasing frequency among immunocompetent individuals.[4] Patients typically present with generalized symptoms such as headache, nausea, vomiting, and confusion.[5] Despite advances in radiologic techniques, the diagnosis of PCNSL remains a challenge. In this article, we describe a case of periventricular PCNSL in an 82-year-old female presenting as a shunt malfunction.

Case report

History and presentation

An 82-year-old female initially presented 5 years ago to an outside hospital with magnetic gait, memory difficulties, and urinary incontinence, consistent with normal pressure hydrocephalus (NPH). She underwent right frontal ventriculoperitoneal (VP) shunt placement, which improved her symptoms (Figure 1(A)). She presented again in June 2017 to an outside hospital with signs consistent with worsening NPH symptoms: worsening gait dysfunction and urinary incontinence without headache or vomiting. Computed tomography (CT) scan at that time did not reveal any abnormalities (Figure 1(B)). Multiple adjustments of her shunt failed to improve her symptoms. The patient ultimately presented to our emergency department several weeks later with worsening magnetic gait, urinary incontinence, and confusion. Patient did not complain of headaches, visual symptoms, or nausea/vomiting. Neurologic examination revealed orientation to self only and sensory disturbances in both the upper and lower limbs. Follow-up CT imaging displayed collapse of the right ventricle (Figure 1(C)). The patient was admitted for revision of shunt with possible septostomy.

Operation

The patient underwent a proximal revision of the right VP shunt with Y connector placement. Worsening collapse of the right ventricle was noted following the procedure (Figure 1(D)). Magnetic resonance imaging (MRI) on postoperative day one revealed extensive subependymal nodular enhancement along the lateral ventricles greater in the right than left, foramen of Monroe, and fourth ventricle along the inferior medullary velum (Figure 2).

Postoperative findings and treatments

CSF fluid was collected from the shunt and sent for cytology, which revealed CD45+ cells concerning for malignancy. A CT scan of the chest was performed due to concern for possible pulmonary embolus and revealed bilateral lobar filling defects which led to IVC filter placement. Moreover, the CT revealed prominent left axillary and submental lymph nodes. These were evaluated, biopsied by otolaryngology, and determined to be negative for malignancy. Because of uncertainty of the pathology, we proceeded with an endoscopic brain biopsy of the lesion within the lateral ventricles (Figure 3). The immediate postoperative period was uneventful. Patient's family consented via telephone to discussion of case details.

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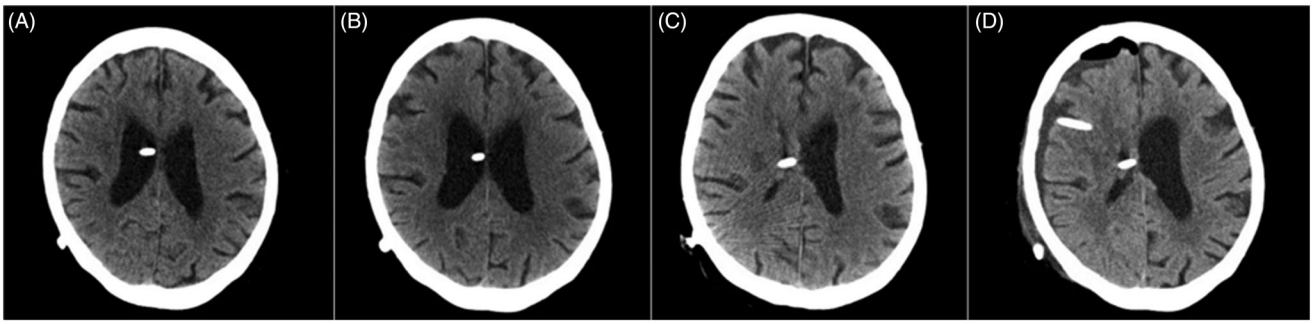

Figure 1. Timeline of axial computed tomography scans. (A) Patient's CT 2 years prior while asymptomatic. (B) CT taken upon presentation to outside hospital (7 weeks prior to admission). (C) CT performed following opening pressure adjustment (1 week prior to admission). (D) CT performed following shunt revision with Y connector placement.

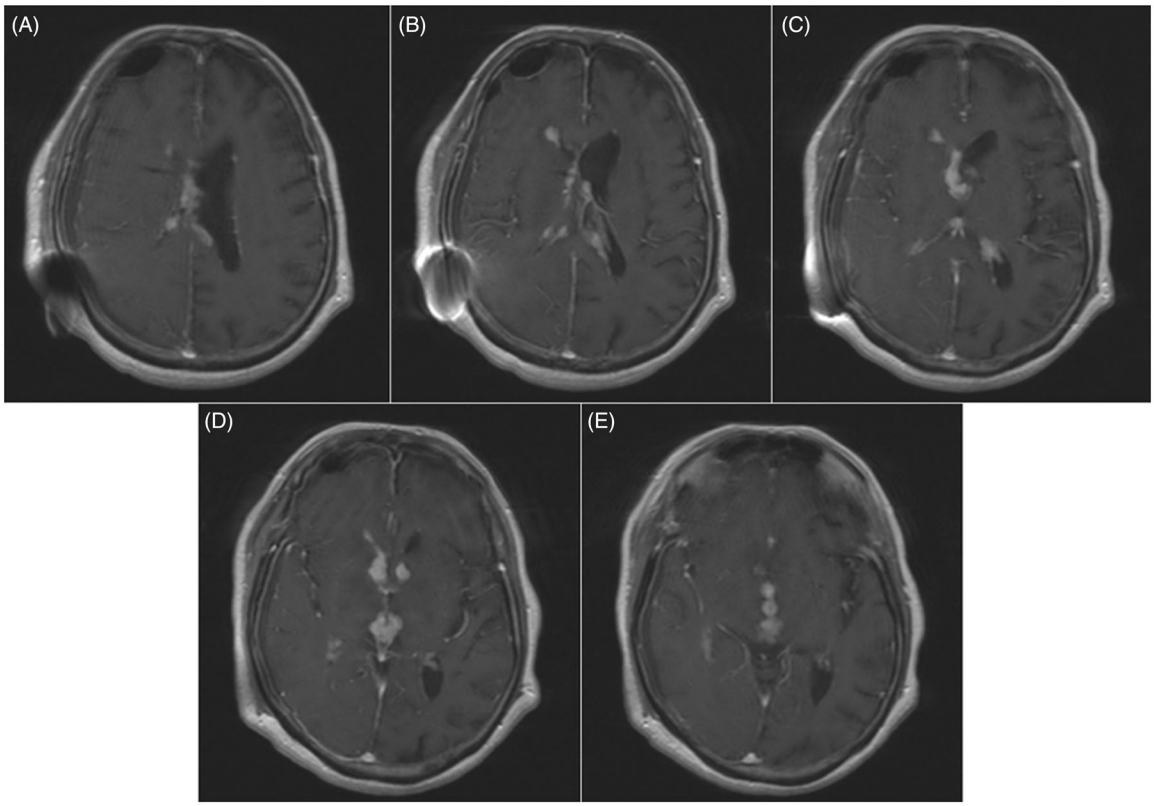

Figure 2. Postoperative axial MRI study (T1 with gadolinium) displaying subependymal nodular enhancement along the ventricular system.

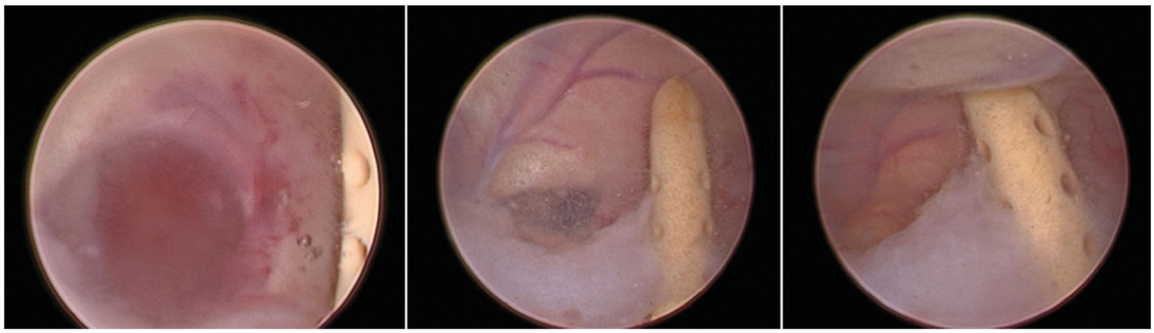

Figure 3. Intraoperative photographs from endoscopic biopsy. (A) Foramen of Monroe to the bottom left and VP shunt catheter on the right side. (B) Shunt catheter with attached grayish lesion. Foramen of Monroe in the bottom left. (C) Close-up of the lesion attached to the catheter.

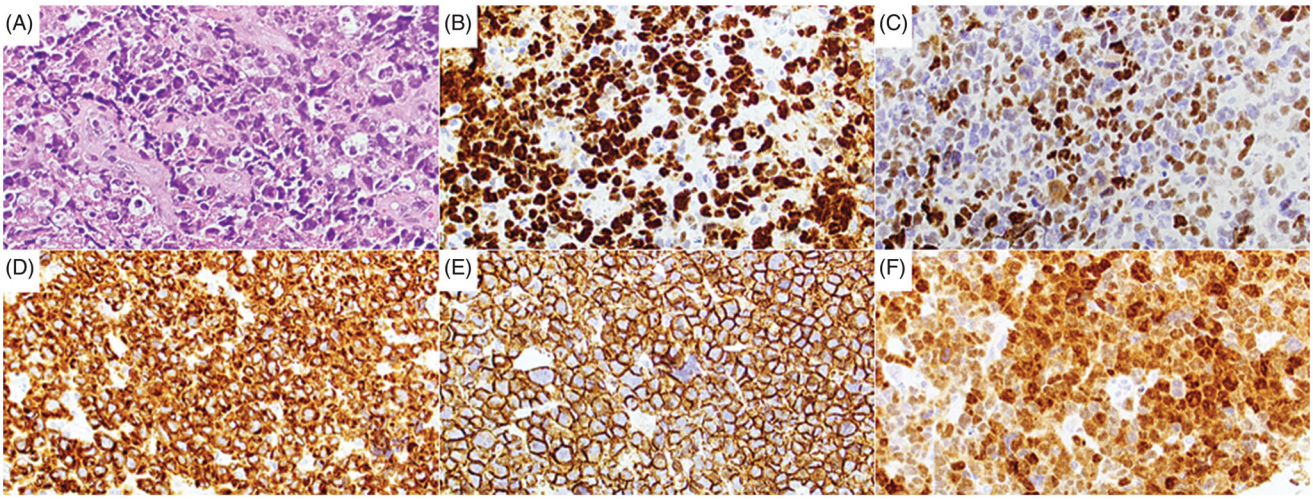

Figure 4. Photomicrographs. (A) H & E staining demonstrating sheets of large anaplastic and pleomorphic cells. (B) Immunostaining with Ki67, a marker for cellular proliferation. (C) Immunostaining with c-myc, an oncogene. (D) Immunostaining for Bcl-2, a regulator protein for apoptosis. (E) Immunostaining for CD20 demonstrating tumor cells with a B-cell immune phenotype. (F) Immunostaining with MUM1, and activation marker.

Histopathological, immunohistochemical, and molecular study

Histopathological examination of the specimen revealed sheets of large anaplastic and pleomorphic tumor cells. Stains were positive for Ki67, c-myc, Bcl-2, CD20, and MUM1. This immune phenotype is compatible with diffuse large B-cell lymphoma (Figure 4). Bone marrow biopsy was performed and was negative. The patient was referred for oncological management of her disease.

Discussion

To our knowledge, this is the first reported case of PCNSL in a patient with NPH treated with a VP shunt presenting as a shunt malfunction. Diffuse large B cell lymphoma is an aggressive tumor and early diagnosis is critical for optimization of therapy and prognosis. Despite advances in imaging, the diagnosis remains difficult. The typical appearance on CT imaging is usually characteristic of solitary intraparenchymal, well-defined, lobulated lesions which typically abut ependymal or meningeal surfaces.[6,7] The lesions appear either isodense or hyperdense to gray matter. There is well-circumscribed homogenous enhancement following the administration of IV contrast.[6] On MRI, less signal intensity on T1 and variable intensity on T2 is typically observed.[1] Lesions are most commonly located supratentorially with isolated lobar involvement.[6]

There are various atypical presentations of primary CNS lymphoma that have increased in recent years. Intraventricular manifestations are very rare, with very few instances documented.[8–10,11,12] Subependymal involvement, as seen in our patient, has been described previously.[6,13] PCNSL lesions presenting as diffuse ventriculitis have also been reported.[2] Ventricular involvement is typically associated with contiguous extension of a parenchymatous lesion.[6,13] Thus, diffuse periventricular enhancement without a mass is uncommon.

Our patient presented atypically with a periventricular pattern of PCNSL. Continued follow up with serial imaging was critical for identifying PCNSL in this patient. The patient's clinical symptoms preceded changes in imaging, making the diagnosis more difficult. It is important to consider alternative diagnoses in the setting of shunt complication with a repeated lack of response to opening pressure modification. The knowledge of this phenomenon is not exclusive to PNCSL but may occur in the setting of high-grade glioma, or other CNS malignancies. However, this rare case of PCNSL presenting as a VP shunt complication highlights the importance of a broad differential diagnosis and multidisciplinary evaluation when treating a patient with signs of shunt malfunction.

One previous case of PCNSL with ventricular involvement presenting as a shunt malfunction has been reported in the literature.[14] The case describes a 22-year-old male with VP shunt similarly presenting with a prolonged course of shunt malfunction refractory to traditional management. However, the patient underwent craniotomy to explore a presumed dilated temporal horn but was found to have subependymal lesions that were subsequently diagnosed at PCNSL. Despite two rounds of chemotherapy, the patient succumbed to his disease 2 months later secondary to seeding of the fourth ventricle, brain stem, and cervical spine. Thus, the case demonstrates that early diagnosis of PCNSL is critical, and initial surgical intervention is typically biopsy, unless urgent decompression is required.[15]

We describe the first published report of an 82-year-old patient with NPH and a VP shunt, who presented with prolonged shunt malfunction refractory to traditional management. The case illustrates the importance of a broad differential in patients with NPH and refractory shunt malfunction.

Disclosure statement

None of the authors or any member of his or her immediate family has funding or commercial associations (i.e. consultancies, stock ownership, equity interest, patent/licensing arrangements, etc.) that might pose a conflict of interest in connection with the submitted article.

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