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Case Report

Unusual presentation of secondary CNS lymphoma with punctate intralesional and intraventricular hemorrhage ☆

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ABSTRACT

Primary and secondary central nervous system lymphomas are infrequently encountered entities that present nonspecifically for which imaging plays a critical role in the diagnostic process. The conventional wisdom is that the presence of intralesional hemorrhage in an immunocompetent patient strongly counts against the diagnosis of lymphoma; however more recent evidence suggests that the distinction is more nuanced. Especially for PCNSL, there is increasing recognition that some degree of hemorrhage occurs more frequently than previously thought, and there are a number of case reports describing various types of hemorrhagic lesions that were histologically confirmed to be lymphoma. Hemorrhage in SCNSL is much rarer with only 3 cases described to date. We describe an unusual case of SCNSL with a pattern and progression of hemorrhage distinct from the prior descriptions. Awareness of the range of possible appearances of atypical presentations of CNS lymphoma is important to ensure that it is not prematurely excluded from the differential diagnosis which can delay appropriate treatment. Although rare, intralesion hemorrhage in itself should not preclude the diagnosis of lymphoma particularly when the other imaging features are congruent.

Introduction

Secondary central nervous system (CNS) lymphoma (SCNSL) which represents CNS spread of systemic lymphoma is one of 2 subtypes of CNS lymphoma, the other being primary CNS lymphoma (PCNSL) where disease is confined to the CNS without systemic involvement [1]. Primary CNS lymphomas although the second most common primary brain malignancy in adults, account for just 6.6% of all malignant brain tumours [2]. SCNSL is more common, seen in up to 27% of non-Hodgkin's lymphoma (NHL) while PCNSL comprises

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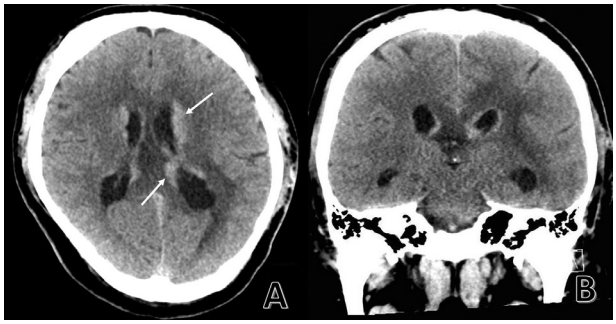

Fig. 1 – Admission noncontrast CT brain showing diffuse nodular hyperdense lesions lining the lateral ventricles (A, solid arrows) with resultant nonobstructive hydrocephalus (B).

just 2%-3% of NHL cases [3]. The presence of intralesional hemorrhage in an immunocompetent patient is considered atypical for lymphoma and historically used as a factor against the diagnosis of lymphoma [2]. Although there is increasing recognition that a certain degree of hemorrhage does not preclude the suspicion of PCNSL, it is still exceedingly rare in SCNSL with only 3 prior cases recorded. Here, a case with imaging features different from the prior ones is reported, and the important radiological implications for diagnosis discussed.

Case presentation

A 46-year-old Asian male with a background of obesity, gallstones and hypertension presented acutely with a few days history of frontal headaches, dizziness, anorexia, gait instability and short term memory loss. His neurological examination was otherwise normal. The initial noncontrasted computed tomography (CT) brain showed diffuse, nodular, hyperdense lesions along the ventricles with nonobstructive hydrocephalus (Fig. 1).

Subsequent magnetic resonance imaging (MRI) demonstrated homogeneously enhancing, T2 w hypointense periventricular and ependymal lesions with moderate restricted diffusion, scattered punctate foci of susceptibility and a trace amount of intraventricular hemorrhage (Fig. 2). The initial impression based on his clinical presentation and imaging features was of ventriculitis for which he received meningitic doses of antibiotics and antivirals.

His admission blood cell counts and serum lactate dehydrogenase were normal. A lumbar puncture revealed an elevated opening and closing pressure, and an elevated white cell count with a lymphocytosis (61%). Cerebrospinal fluid cultures and polymerase chain reaction were clean. Further evaluation with a CT thorax, abdomen and pelvis, autoimmune panel and HIV screen were all unrevealing. The patient progressively worsened, becoming increasing confused and drowsy. A repeat MRI for surgical planning found an increase in extent and susceptibility of the lesions (Fig. 3).

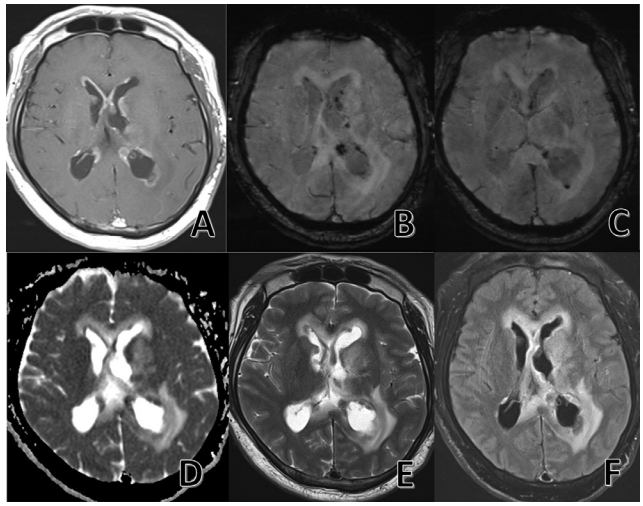

Fig. 2 – MRI demonstrated homogenously enhancing periventricular and ependymal/subependymal lesions (A) with scattered punctate foci of internal susceptibility (B) and sliver of intraventricular hemorrhage (C, solid arrow). The lesions showed moderate restricted diffusion (D), and were T2w-hypointense (E) with periventricular white matter hyperintensities likely a combination of vasogenic and transpendymal oedema (E, F). No parenchymal enhancing lesion or leptomeningeal enhancement was seen.

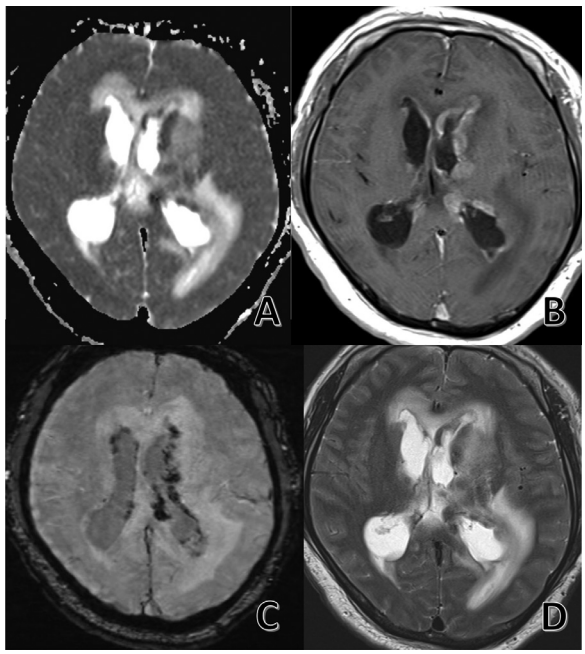

Fig. 3 – Preoperative MRI 5-days after the initial scan revealed progression of the lesions (A, B) with increased haemorrhage (C) and greater mass effect and cerebral oedema (D).

Table 1 – Summary of reported cases of intratumoural hemorrhage associated with SCNSL.

Author, date	Age	Gender	Known systemic lymphoma	Presentation	Distribution of lesions	Pattern of hemorrhage	Subtype
Demir et al., 2017 [8]	72	Female	No	Altered mental status, right hemiparesis	Parenchymal, single lesion in left frontal lobe	Hemorrhagic mass	Not specified
Suzuki et al., 2017 [9]	76	Male	Yes, B-cell lymphoma	Altered mental status, seizures	Parenchymal; single lesion in left temporal lobe	Hemorrhagic mass	Intravascular lymphoma
Malivoka et al., 2018 [10]	74	Male	No	Altered mental status	Parenchymal; multiple lesions in grey/white matter junction and corpus callosum	Discrete hemorrhagic foci	Intravascular lymphoma

The patient underwent stereotactic biopsy of the left periventricular lesion and insertion of an external ventricular drain 6 days after admission. An immediate postoperative CT scan showed a small amount of intraventricular blood with worsening generalized oedema and mass effect. The histology returned as diffuse large B-cell lymphoma with immunohistochemistry stains positive for CD20, CD79a, BCL-2, BCL-6, MUM1 and myc. CD10 was negative. Dexamethasone 8 mg BD was started immediately after biopsy without improvement in the patient's condition. He was transferred to a tertiary institution for further management where he was initially treated for presumptive PCNSL. A bone marrow trephine was performed 1 day after transfer and immunohistochemistry stains for CD20 and CD79a highlighted a small number of scattered and singly occurring large B lymphocytes that were not readily apparent on the hematoxylin and eosin slide, suspicious for low-level involvement by diffuse large B-cell lymphoma. After discussion at the lymphoma multi-disciplinary tumor board meeting, a diagnosis of diffuse large B-cell lymphoma with secondary CNS involvement was established and the treatment correspondingly modified with good treatment response. A PET/CT performed 5 months after initiation of treatment revealed no suspicious FDG avid nodal or extranodal disease.

Discussion

Classically, SCNSL is said to have a predilection for the leptomeninges, with involvement seen in two-thirds of patients and parenchymal lesions seen in the other one-third [1]. More recent data from large clinical trials suggests that parenchymal involvement is more common, seen in 40%-60% of patients, while leptomeningeal spread occurs in 20%-30% and 10% have disease at both sites [2]. The higher proportion of parenchymal involvement is an important radiologic consideration since the distribution of disease is less useful for differentiating SCNSL from PCNSL [2]. Exclusive leptomeningeal or ependymal/subependymal lesions, as was seen in our case, should still prompt the suspicion for SCNSL over PCNSL for which parenchymal lesions are almost constant [2]. Poor clinical or radiological response to steroids could also help favor SCNSL over PCNSL which typically responds dramatically to steroids.

There is increasing recognition that intratumoural hemorrhage occurs more frequently in PCNSL than previously thought. Descriptions from several case reports include: (a) a cortical pattern [4] (b) lesions with hemorrhagic components [5] (c) a hemorrhagic mass [6], and (d) appearing as pure hematomas [7]; a recent retrospective review of 19 cases found intratumoural foci of susceptibility on susceptibility weighted imaging (SWI) in 10 patients (53 %) and gross tumoral hemorrhage as seen on T2-weighted imaging in 4 (21 %) [5].

Intratumoural hemorrhage with SCNSL is much rarer; in a recent retrospective review of 21 patients, hemorrhage was found in only one [8], and only 3 cases have been reported thus far [8–10]. In all 3 cases, the patients were elderly and the lesions were parenchymal. Two cases appeared as hemorrhagic masses while the last demonstrated small discrete areas of hemorrhage within the lesions; associated mass effect was present in all. Two of the 3 cases were of the intravascular lymphoma subtype. The cases are summarized in Table 1.

In our case, the patient was much younger, and his imaging features differed from the previous descriptions: firstly, the lesions were diffuse, with a periventricular and ependymal/subependymal distribution; secondly, the hemorrhage appeared as scattered punctate foci within the lesion without associated mass effect along with intraventricular extension. Rapid progression of the hemorrhagic components was observed, which also bled intra-operatively.

Conclusion

Atypical presentations of CNS lymphoma are infrequently encountered and awareness of the range of possible appearances is critical to ensure that it is not prematurely excluded from the differential diagnosis. Although rare, the presence of hemorrhage does not automatically preclude the possibility of SCNSL, particularly when other imaging features are supportive.

This case highlights that the morphology of hemorrhage can be protean; rapid progression of the hemorrhagic components and intraoperative lesional bleeding can also occur. This pattern of hemorrhage in SCNSL is different from the 2 types previously reported and we hope it will add to the diagnostic armamentarium in dealing with similar cases in future.

Patient consent

Informed consent was obtained from the patient for publication of this case report and accompanying images.

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