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A rare case of suspected intravascular large B-cell lymphoma forming nodular masses in the brain and around the prostate and seminal vesicle

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ABSTRACT

Intravascular large B-cell lymphoma (IVLBCL) is a rare subtype of diffuse large B-cell lymphoma (DLBCL). Furthermore, tumorigenesis is extremely rare. An 80-year-old man was admitted to our hospital with nervous symptoms. Imaging tests showed a brain tumor and mass lesions in the seminal vesicle and retroperitoneum. Transrectal biopsy of the seminal vesicle helped diagnose the patient with DLBCL. The patient's general status deteriorated rapidly, and he died on the 23rd day after admission. An autopsy was performed and the pathological diagnosis was DLBCL, specifically suspected as IVLBCL, with nodular masses in the brain and seminal vesicle.

1. Introduction

Extranodal lymphoma accounts for 30–50% of non-Hodgkin lymphoma, and most of the cases are of the diffuse large B-cell lymphoma (DLBCL) type.[1] In the past, intravascular large B-cell lymphoma (IVLBCL) was defined as a rare subtype of DLBCL whose incidence was less than 1%. Recently, IVLBCL has been considered as an independent type of extranodal lymphoma by the World Health Organization (WHO) classification, which is different from one of the subtypes of DLBCL.[1]

Because IVLBCL rarely forms nodular masses, it is difficult to diagnose at the early phase of disease. While in the past a lot of patients were diagnosed by performing autopsy, IVLBCL can now be diagnosed at an early stage as a result of developments in the knowledge of their pathogenesis and methods of examination. The pathological characteristic of IVLBCL is selective growth of lymphoma cells within the lumina of vessels. In previous studies, it is shown that invasion of tumor cells often occurs at the bone marrow, liver, spleen, skin, or lung.[2] Some cases present several symptoms when lymphomatous cells occlude the lumina of small vessels of any organ. If the patients show some specific symptoms, a biopsy on the affected site may help in the early diagnosis of IVLBCL.

IVLBCL with organ infiltration often shows diffuse visceral invasion upon examination by imaging, rarely forming nodular masses. There are few case reports on IVLBCL forming nodular masses.[3–5] To the best of our knowledge, there are no reports of cases with nodular masses formed by IVLBCL in the genitourinary organs. Here, we report the first case of lymphoma suspected of IVLBCL diagnosed by autopsy with the formation of several nodular masses in the seminal vesicle, prostate, brain, and retroperitoneal lymph nodes.

2. Case presentation

An 80-year-old Japanese man visited our hospital showing signs of inarticulation. He had no significant past or family history of cancer. Physical examination revealed impaired walking and a positive Barré's sign in left arm.

Left hydronephrosis and mass lesions around the seminal vesicle, prostate, and retroperitoneum were found by computed tomography (CT) scan (Fig. 1). He did not have hepatomegaly nor splenomegaly. The interleukin-2 (IL-2) receptor value was high and there was no cytopenia during blood test (Table 1). Magnetic resonance imaging (MRI) enhanced by gadolinium showed two

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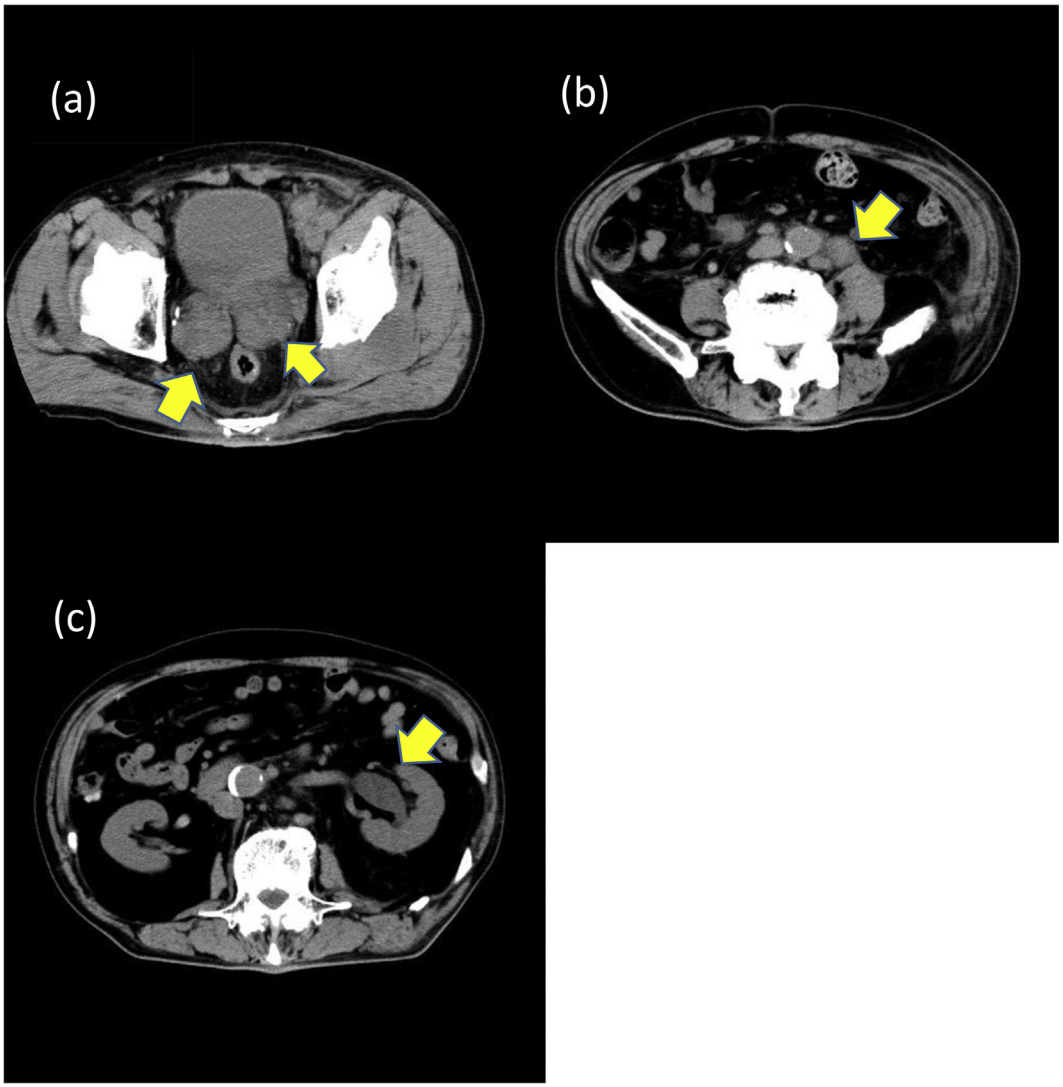

Fig. 1. (a): Seminal vesicle was bilaterally enlarged. (b):Computed tomography shows mass lesion of retroperitoneum. (c): Computed tomography shows left hydronephrosis.

nodular masses with inflammation in the brain. Then, a fine-needle aspiration biopsy of the seminal vesicle and prostate was performed. There were a lot of atypical large lymphocytes. The results for CD20 and CD79 were positive, while those for CD3 and CD5 were negative. From these findings, DLBCL originating from the seminal vesicle was suspected. As his status was poor, he and his family opted for the best supportive care. Dexamethasone of 2 mg/dose was administered for edema and inflammation resulting from brain tumor. He died 23 days from hospitalization, and the autopsy was performed at our institution. Atypical lymphocyte infiltrates each organ, especially the seminal vesicle and brain. For the detection of the nature of the tumor, immunohistochemical staining was done. The results for CD20 and CD79 were positive, while those for CD3 and CD5 were negative (Fig. 2). There were atypical lymphocytes proliferation within the lumina of small vessels of these organs. Furthermore, there were similar findings in the mesentery, pancreas, and lymph node around the aorta. Based on the autopsy, he was diagnosed with DLBCL suspected of IVLBCL infiltrating into the prostate, bladder, lung, liver, gallbladder, spleen, stomach, adrenal and pituitary glands, bone marrow, kidney, mesentery, pancreas, and lymph node around the aorta with nodular mass lesions in the brain, prostate, and seminal vesicle.

3. Discussion

IVLBCL is regarded as an aggressive lymphoma and therefore has a poor prognosis. Due to the absence of the formation of mass lesions, the diagnosis of IVLBCL is often delayed. The prognosis and response to chemotherapeutic treatment of IVLBCL are not clarified since there have been no large-scale prospective clinical trials. As a retrospective data, Shimada et al. reported that the 2-year survival rate of IVLBCL patients who underwent R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone) treatment is 66%, and the treatment plan of IVLBCL is equivalent to those of other lymphomas. Although IVLBCL has a rare incidence itself and is difficult to diagnose, large-scale randomized controlled trials are expected in future works.

IVLBCL is a subtype of extranodal lymphoma and does not usually show tumor formation and lymphadenopathy except for lymphoma cells within the lumina of small vessels in various organ systems.

There have been several previous reports on IVLBCL infiltrating the bone marrow, skin, liver, or lung. However, this case of IVLBCL with nodular masses is, to our best knowledge, the first of its kind with a tumor in the adrenal gland.[2] In our case, the autopsy showed