AV nodal dissociation and facial droop: An atypical presentation of lymphoma

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Introduction

Diffuse large B cell lymphoma is the most common lymphoid malignancy. [1]

Symptoms commonly consist of lymphadenopathy and constitutional symptoms at diagnosis.

Extra-nodal symptoms occur in 20-40% patients at initial diagnosis.

Cardiac conduction abnormalities and neurologic deficits are rarely the presenting symptoms of DLBCL. [2]

Case

50-year-old female with initial presentation of shortness of breath and chest pain was found to be bradycardic with hypotension (HR: 40, BP: 92/51).

She was treated in an ICU with atropine injections and a dopamine infusion for symptomatic bradycardia.

Electrocardiogram revealed a junctional bradycardia (HR: 40). Ischemic workup was pursued for lower extremity swelling, dyspnea on exertion, and elevated troponin (46ng/L). Transthoracic echocardiogram revealed a normal ejection fraction (60%) and no valvular abnormalities. A left heart catheterization was unrevealing.

Additionally, the patient had an associated left sided facial droop that developed early during her clinical course. Differential included Lyme disease or Ramsay Hunt Syndrome. Head CT showed no evidence of intracranial abnormality.

Case (cont.)

Given her presentation of symptomatic bradycardia, Lyme disease was suspected. She was treated with ceftriaxone and later pacemaker placement after Lyme titers returned negative. She was also started on antivirals for facial droop due to concern for Ramsay Hunt Syndrome. With negative titers and neurologic abnormalities, a more systemic disease was considered.

A left renal lesion was identified after performing a CT scan of her pelvis for leg pain. Described as area of enhancement concerning for infiltrative malignancy, it was biopsied.

Her presenting symptoms continued, including lower extremity edema while awaiting biopsy results and she was again admitted. She also developed atrial flutter.

Labs & Imaging

WBC: 26k/uL **Hgb**: 10.9g/dL **Plt**: 198k/uL
Differential: neutrophilia

Peripheral smear: no blasts
Lyme titers: negative

Lactate: 2.1 **CRP**: 4.52 **ESR**: 10 **LDH**: 778

ALT: 342 **AST**: 176 **ALP**:86 **Tbili**: 0.4

CT body: Enhancing densities most significant at lateral aspect of the left kidney measuring 2.5cm thick with bilateral urethral thickening. Findings consistent with infiltrative malignancy.

Left renal lesion biopsy: aggressive B-cell lymphoma

FISH: high-grade B-cell lymphoma; double hit (C-Myc, BCL-6 rearrangements on FISH)

CSF: Lymphoma cells.

Bone marrow: Negative for lymphoma.

PET/CT: Consistent with bony lesions, pericardial disease and abdominal organ infiltration

MRI: leptomeningeal & cranial nerve enhancement of CNS and spinal cord consistent with CNS involvement

Clinical Course

Left kidney biopsy **revealed high-grade B-cell double-hit DLBCL**, and treatment was initiated during her hospitalization.

Ramp-up therapy: R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, prednisone)

Definitive induction therapy: DA-R-EPOCH (rituximab, etoposide, prednisone, vincristine, cyclophosphamide, doxorubicin)

CNS prophylaxis: via Ommaya reservoir: etoposide, cytarabine, methotrexate, gemcitabine, rituximab.

Despite optimal treatment, her DLBCL progressed, and she experienced complications including pain related to osteolytic lesions, Ommaya reservoir infection, adrenal insufficiency, and debilitating neurologic symptoms including cranial deficits, dysphagia, recurrent aspiration, headaches, and neuropathic pain.

Conclusions

Cardiac conduction and cranial nerve abnormalities are a marker of late and aggressive disease if associated with DLBCL. [3]

Cardiac conduction abnormalities as a marker of CNS involvement is a unique presentation of DLBCL.

Myocardial involvement is more often asymptomatic and identified on autopsy.[1,4]

Cardiac MRI is the best test to identify specific lesions of the cardiac conduction system. [3]

Maintain a broad differential diagnosis. This patient's signs and symptoms are a marker of systemic disease resulting in a diagnosis of a high-grade lymphoma.

References

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