

Cutaneous metastasis of primary diffuse large B-cell lymphoma of the central nervous system

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Background

- Primary **diffuse large B-cell lymphoma (DLBCL)** of the **central nervous system (CNS)** is a rare but aggressive variant of extranodal non-Hodgkin lymphoma that represents 3-4% of all primary brain tumors.
- Primary DLBCLs of the CNS have a high propensity to relapse locally within the CNS, whereas only a few cases of **cutaneous metastasis** have been described in the literature.

Case Report

- We present a unique case of cutaneous metastasis of a primary DLBCL of the CNS in a 79-year-old man who was in clinical remission for four years until presenting with recurrence in the left adrenal gland and several nodular lesions on the left flank (Figure 1A-B).
- Skin biopsy revealed a diffuse dermal infiltrate of atypical B-cell lymphocytes with expression of CD20, BCL-2, BCL-6, and MUM-1, consistent with DLBCL (Figure 2A-F).
- Immunoglobulin heavy chain (IgH) gene rearrangement demonstrated that the DLBCL of the skin was of the same clonal origin as the primary DLBCL of the CNS.
- The patient has completed three cycles of rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP) chemotherapy and daily ibrutinib, with plans to complete a total of six cycles.
- Restaging CT three months after initiating treatment showed complete resolution of the left adrenal mass and resolving nodularity of the skin lesions on physical exam.

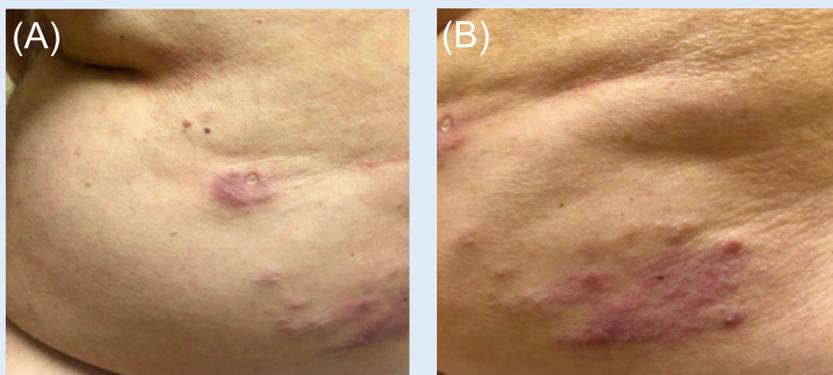


Figure 1.

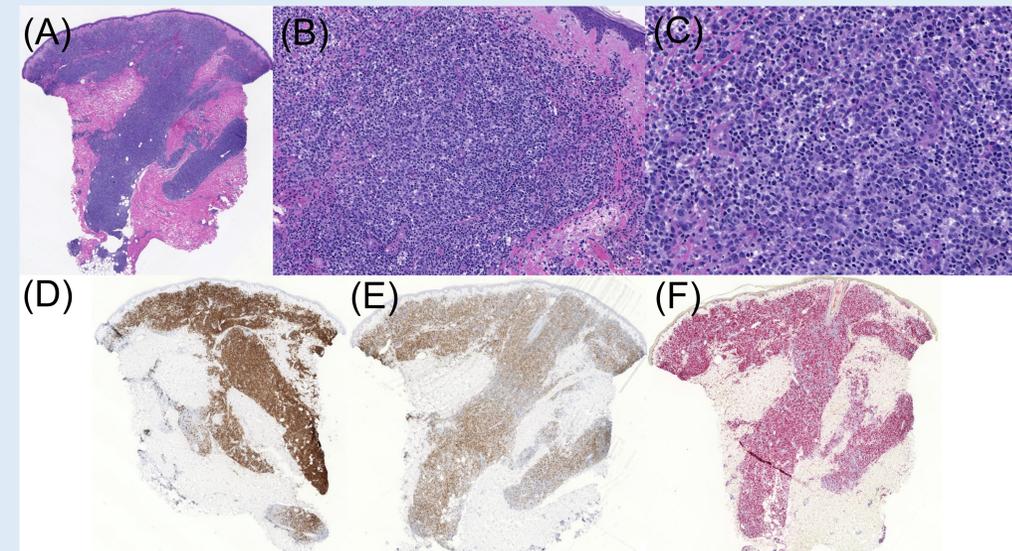


Figure 2.
A-C) H&E
D) CD20
E) MUM-1
F) Ki-67

Discussion

- Patients with a history of primary DLBCL of the CNS in long-standing remission who present with new, atypical lymphoid infiltrate represent a **diagnostic challenge** in determining whether this is metastasis of the primary DLBCL of the CNS or a primary cutaneous DLBCL.
- **Gene rearrangement studies** comparing IgH clones of both the primary DLBCL of the CNS and the cutaneous nodules allowed us to be certain that this represented cutaneous metastasis of the patient's primary DLBCL of the CNS.
- The patient we describe is unique from other documented cases in that his cutaneous metastasis presented significantly later after the initial diagnosis of primary DLBCL of the CNS.
- Our patient is also the first to present with **evidence of distant metastasis** in the adrenal gland at the time of diagnosis of the cutaneous metastasis.
- To date, only two patients have been reported to achieve clinical remission of the cutaneous metastasis and these patients were treated with either radiotherapy or rituximab plus cyclophosphamide, vincristine, and prednisone chemotherapy.
- We present the **first case** in which clinical improvement of cutaneous metastasis was achieved with ibrutinib plus R-CHOP chemotherapy.

Conclusion

- **Cutaneous metastases from primary CNS lymphomas** are rarely observed, and as presented in our case, may occur **long after remission** is achieved. Utilization of **gene rearrangement studies** can help discern the relationship between the primary and secondary lesions.