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Case report/ Kazuistyka

Precursor B-lymphoblastic lymphoma mimicking: An acute subdural hematoma

Ant Uzay [1,*], *Siret Ratip* [1], *Ilhan Elmaci* [2], *Metin Ozdemirli* [3]

[1] *Department of Haematology, Acıbadem Hospital, Istanbul, Turkey*
[2] *Department of Neurosurgery, Memorial Hospitals Group, Istanbul, Turkey*
[3] *Department of Pathology, Georgetown University Hospital, Washington D.C., USA*

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ABSTRACT

Objective and importance: We present the first case of a precursor acute subdural B-lymphoblastic lymphoma mimicking an acute subdural hematoma. **Clinical presentation:** A 19 year old male presented with an acute onset of headache, nausea and vomiting. CT scan showed crescentic right-sided, frontoparietal subdural mass isointense with cortex and showing homogeneous enhancement after gadolinium. **Intervention:** The patient underwent a craniotomy and a gray subdural tumor with invasion of both dura and brain was observed. The invaded dura was resected and duraplasty performed. Histopathologically, the tumor was composed of small round cells infiltrating soft tissue. In some areas of the tumor, cells were arranged in a linear, "Indian file" fashion between collagen bundles. Their nuclei were generally uniform, round to ovoid in shape, small to medium in size, and featured delicate chromatin. Accompanying cytoplasm was scant. Necrosis was absent. On immunohistochemical analysis, the tumor cells were positive for CD79a, TdT, CD10 and CD34. **Conclusion:** Subdural lymphoma can present as a neurosurgical emergency, and lymphoma should be considered as a rare but possible diagnosis before operation.

Introduction

Presentation of patients with solitary involvement of the dura by lymphoma is rare and is typically that of a subacute to chronic process. We present the first case of a patient with precursor B-lymphoblastic lymphoma mimicking an acute subdural hematoma.

Case report

A 19-year-old male patient was referred to Marmara University Hospital from a peripheral hospital following unsuccessful burr-hole drainage of a subdural hematoma. Presenting complaints included severe headache, nausea, and vomiting. A physical examination revealed papilledema

* *Corresponding author at*: Acıbadem University Hematology, Betül sokak, Çesan sitesi, 2 Blok, D8, Acıbadem mahallesi, Kadıköy, Istanbul, Turkey. Tel.: +90 5365123022; mobile: +90 5365123022.
E-mail addresses: teteantuzay@yahoo.com, ant.uzay@acibadem.com.tr (A. Uzay).
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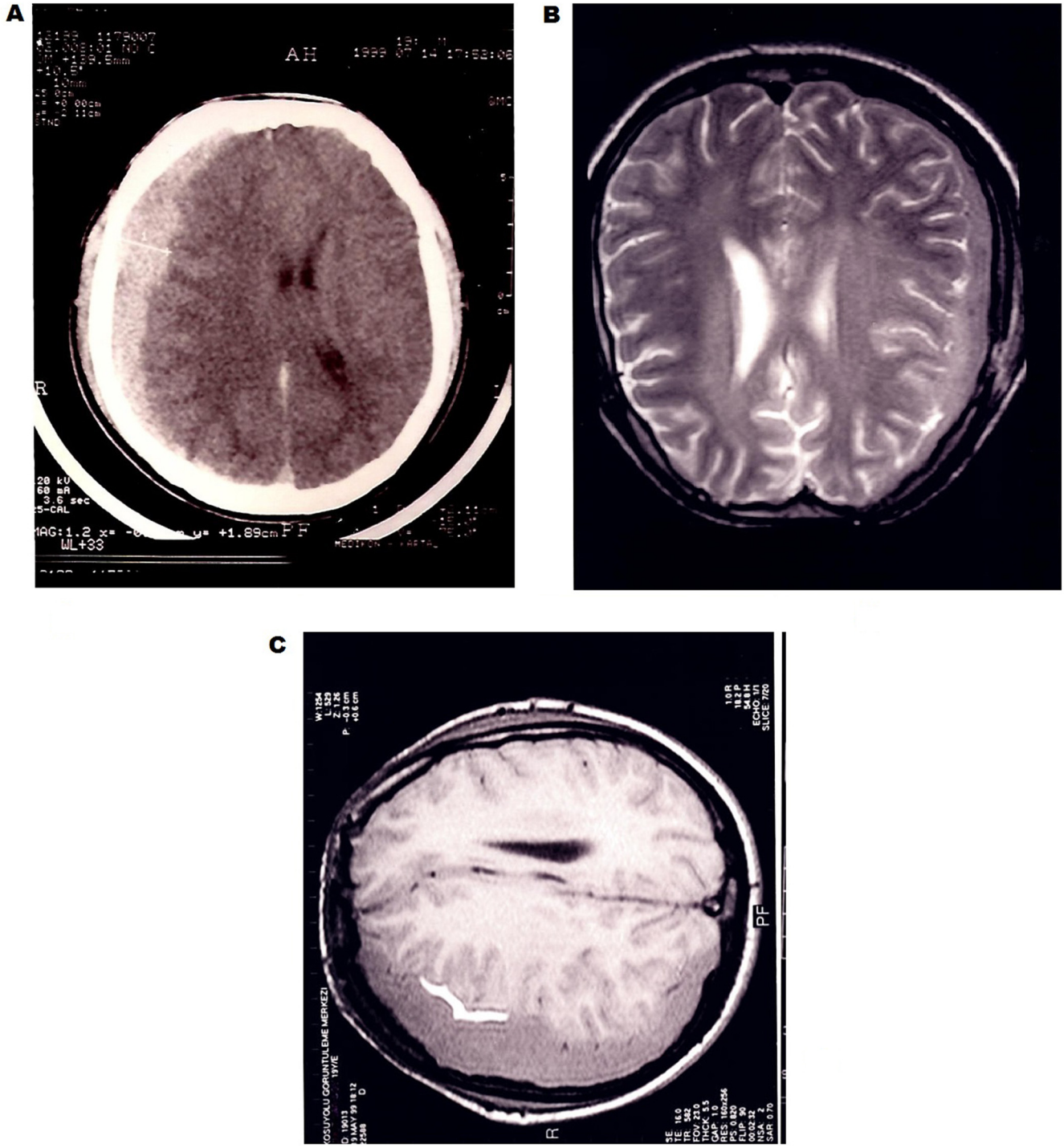

Fig. 1 – A: Axial CT scan. A hyperdense, subdural mass in the right frontoparietal region causing midline shift and compression of the lateral ventricle. B, C: Axial T1- and T2-weighted MR images show the mass to be isointense with cortex

but no other neurological signs. A CT scan showed crescentic right-sided, frontoparietal subdural mass which had the appearance of a subdural hematoma (Fig. 1A). The lesion was isointense with cortex on T1-weighted and T2-weighted MR examinations and showed homogeneous enhancement after gadolinium (Fig. 1B, C). The patient underwent a craniotomy and a gray subdural tumor featuring hemorrhage with invasion of both dura and brain was observed. The invaded dura was resected and duraplasty was performed. The postoperative course was uneventful. The formalin-fixed tissue was routinely processed for histologic examination. Immunohistochemical staining was performed using the peroxidase antiperoxidase technique and obtained from Dako Corp, Carpinteria, CA, and antibodies directed against CD3, CD20, CD79a, CD10, CD34, TdT, myeloperoxidase, lysozyme, synaptophysin, GFAP. Histopathologically, the tumor was composed of small round cells infiltrating soft tissue without a specific pattern. In some areas of the

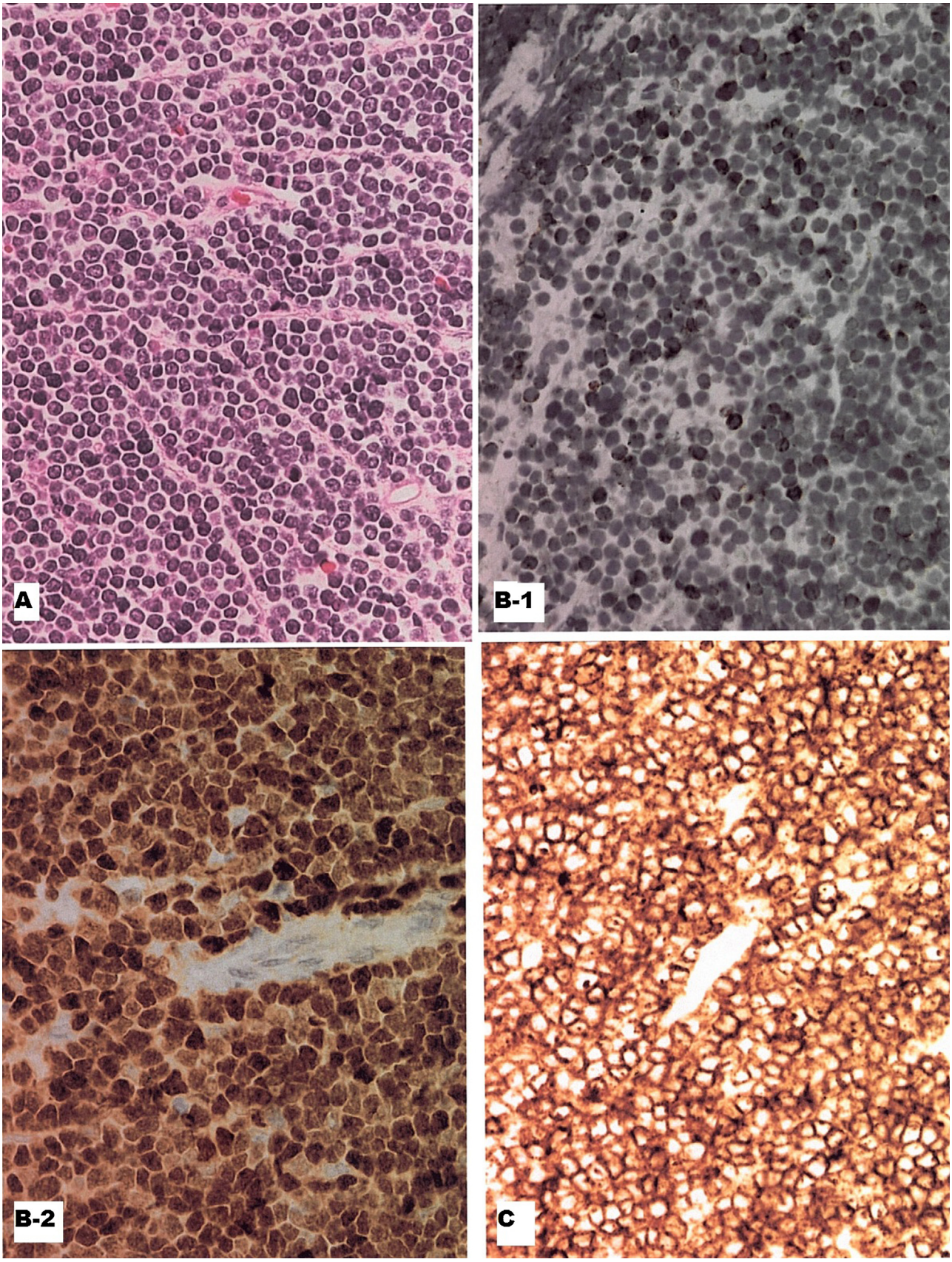

Fig. 2 – Hematoxylene and eosine stain of the brain biopsy, revealing diffuse infiltration of the uniform large immature lymphoid cells (A). The neoplastic cells are immunopositive for TdT (B-1. Positive 79a stain of the specimen; B-2. Strong positive TdT staining of the specimen) and CD10 (C). (A, B and C, ×250)

tumor, cells were arranged in a linear, "Indian file" fashion between collagen bundles (Fig. 2A). Their nuclei were generally uniform, round to ovoid in shape, small to medium in size, and featured delicate chromatin. Accompanying cytoplasm was scant. Necrosis was absent. On immunohistochemical analysis, the tumor cells were positive for CD79a, TdT (Fig. 2B), CD10 (Fig. 2C) and CD34; CD3, myeloperoxidase, lysozyme, GFAP, synaptophysin, and CD20 preparations were negative. A diagnosis of precursor B-lymphoblastic lymphoma was made based on combined cytologic and immunohistochemical findings.

Further assessment of the patient's hematologic status was undertaken. The patient had no B symptoms such as weight loss of greater than 10% body weight, fever or night sweats. The physical examination revealed neither lymphadenopathy nor hepatosplenomegaly. Pertinent laboratory values included a hemoglobin of 14.4 g/dL, a leukocyte count of 7800/mm^3, and a platelet count of 287 000/mm^3. The erythrocyte sedimentation rate was 5 mm/h. Liver function tests and LDH levels were normal as was the peripheral blood smear. The patient was referred to the hematology department. Bilateral posterior iliac crest bone marrow aspirates and trephines were performed. No involvement by lymphoma was detected. A CT scan of the thorax and abdomen showed no sign of lymphadenopathy or hepatosplenomegaly. A diagnosis of primary precursor B-lymphoblastic lymphoma of the subdural space and brain was made. The patient was transferred to the care of the hematology service but, unfortunately, failed to keep outpatient clinic appointments and could not be contacted. He presented two months thereafter with complaints of weakness and lethargy. A full blood count revealed a Hb of 8.1 g/dL, a leukocyte count of 4800/mm^3, and a platelet count of 54 000/mm^3. A repeat bone marrow aspirate and trephine showed heavy infiltration by lymphoblasts. He underwent cranial irradiation, and intrathecal as well as systemic chemotherapy as part of the UKALL X regime, including L-asparaginase, vincristine, daunorubicine, prednisolone, cytarabine, methotrexate, 6-mercaptopurine, thioguanine, cyclophosphamide and etoposide. Despite an initial favorable response, he relapsed at 11 months after onset of treatment and died of the disease.

Discussion

Central nervous system (CNS) involvement by non-Hodgkin's lymphoma (NHL) is usually encountered at advanced stages of the disease [1, 2]. Primary involvement of the CNS by lymphoma remains relatively uncommon [3], and presentation with dural involvement alone is rare [4–11].

With the single exception of an extradural malignant lymphoma of mixed type presenting acutely as a surgical emergency [11], the presentation of patients with solitary involvement of the dura by lymphoma is typically that of a subacute to chronic process [4–10]. The reason for the lack of acute presentations is explained by the low-grade nature of NHLs involving the dura. These include MALT lymphomas [4, 5], small lymphocytic lymphomas [6–8], and a T-cell rich B-cell lymphoma [9].

Lymphoblastic lymphoma as reported herein is a very aggressive form of NHL that usually presents with rapidly progressive lymphadenopathy and shows a high frequency of CNS involvement [3]. Its subtype, B-LBL is uncommon and accounts for less than 10% of cases of lymphoblastic lymphoma [12]. B-LBL has the feature of involving extranodal sites, most often the skin and the bones. In a report of B-LBL, there was no evidence of bone marrow disease at the time of diagnosis in 23 out of 25 patients, and the primary sites of disease were skin (9 cases), bones (5 cases), soft tissue (4 cases), lymph nodes (3 cases), breast (2 cases), stomach and colon (1 case), and mediastinum (1 case) [12]. The patient in this case is the first report of B-LBL presenting with dural involvement. Presentation of lymphoblastic lymphoma, or any other form of NHL, with acute subdural hematoma related to a solitary lesion has also not been previously described. Rapid tumor growth, a reflection of the very aggressive nature of lymphoblastic lymphoma seems to explain the "acute subdural" presentation of this patient.

Even modern neuroimaging methods, such as CT and MRI scans, usually fail to distinguish hematoma from bleeding into tumor at the subdural space [10, 11]. Proton density MRI scans may be suggestive of lymphoma as a hyperintense image and allow differentiation of hematoma and lymphoid tissue [13]. However, when dural lymphoma is associated with hematoma, even proton density MRI sequences are unlikely to differentiate lymphoid tissue from the hematoma. Precursor B-Lymphoblastic lymphoma is a rare subtype of lymphoma, and thus a rare cause of subdural hemorrhage, but tumors associated with haematomas are not rare in neurosurgical practice. Therefore, it is always important to have a differential diagnosis in mind, even for what appears to be a straightforward case, and to always be as prepared as possible for unexpected intraoperative findings.

In the present case, an apparently primary subdural lymphoma presented with acute neurological symptoms unaccompanied by systemic manifestations. Despite investigations, including CT scans as well as bilateral bone marrow aspirates and trephine biopsies, no other site of involvement was identified at the time of presentation. Blood counts and biochemical parameters were also normal at this stage. Nonetheless, two months thereafter, the patient presented with bone marrow involvement. It is possible, of course, that a systemic focus of systemic lymphoma might initially have escaped detection and led to hematogeneous involvement of the dura. Although the pathogenesis of dural involvement by lymphomas is uncertain, most patients with CNS NHL do have bone marrow involvement [14]. Thus, intracranial disease at presentation is usually the result of hematogeneous dissemination. However, as in the present case, manifest marrow infiltration is not a prerequisite for CNS dissemination [1].

Conclusion

Lymphoblastic lymphoma mimicking an acute subdural hematoma and requiring emergency surgical decompression has not been previously reported. The present case indicates

that subdural lymphoma can present as a neurosurgical emergency. Thus, lymphoma should be considered as a rare but possible diagnosis before operation.

Authors' contributions/ Wkład autorów

AU – study design, data collection, manuscript preparation, literature search. SR – data collection, manuscript preparation, literature search. İE – data collection and interpretation. MÖ – data interpretation.

Conflict of interest/ Konflikt interesu

None declared.

Financial support/ Finansowanie

None declared.

Ethics/ Etyka

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; EU Directive 2010/63/EU for animal experiments; Uniform Requirements for manuscripts submitted to Biomedical journals.

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