

Headache, Bilateral Abducens and Incomplete Oculomotor Paresis as the Initial Symptoms in Recurrent Lymphoblastic T-Cell Leukemia – A Case Report

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Abstract

Cranial nerve (CN) involvement in leukemia is rare, usually caused by leptomeningeal spread. The facial nerve and the oculomotor nerves are the most frequently involved CN lesions. Solid depositions of leukemia are rare, and can occur without CNS involvement. A case of a patient with Lymphoblastic T-Cell leukemia (T-LBL) in remission, presented with severe headache and oculomotor dysfunction, due to bilateral infiltration of the cavernous sinus. Despite radiotherapy, clinical deterioration could not be stopped and local pain could not be ameliorated.

Key words: Leukemia; Relapse; Sinus Cavernosus; Cranial Nerves

Introduction

Lymphoblastic T-Cell leukemia (T-LBL) occurs most frequently in young males presenting with lymphadenopathy or mediastinal masses. Less common are extranodal manifestations or abdominal involvement. Complete remission rate is achieved in up to 90% however relapse/progression occurs in approximately 10%.

Cranial nerve (CN) involvement in leukemia is rare, and often caused by leptomeningeal spread. The facial nerve and the oculomotor nerves are most frequently involved. CN lesions due to focal deposits, myelosarcoma or chloromas [1] are rare, and also the infiltration of circumscribed structures as the cavernous sinus has been rarely observed. This case report illustrates the clinical, diagnostic and therapeutic issues in a patient with bilateral infiltration of the cavernous sinus, presenting with pain and oculomotor symptoms.

Case Report

A 29 year old patient with a history of T-LBL since 2014 received an initial chemotherapy [2]. Full remission following consolidation therapy (GM-ALL protocol) for over a year including several cycles of prophylactic intrathecal cytarabine therapy was achieved.

Flow cytometry and bone marrow biopsy (BMB) showed sustained remission and the CSF studies were negative. Following the remission eight month later severe hemicranial headache, bilateral ptosis and an incomplete oculomotor palsy occurred. Magnetic resonance imaging (MRI) showed an infiltrative process

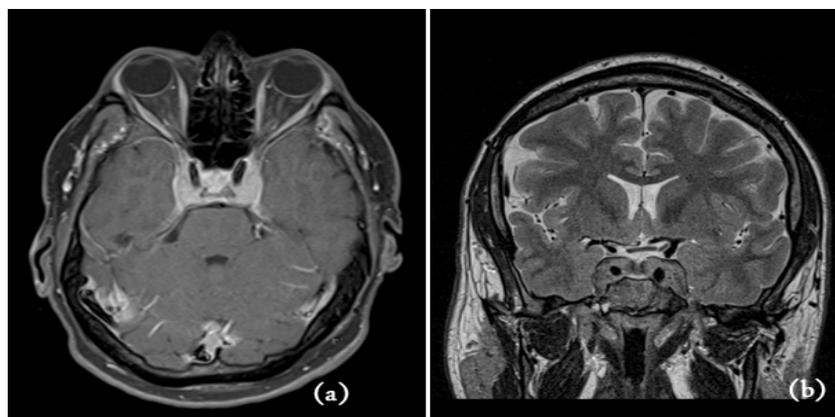


Figure 1: Figure 1a shows thickening of both lateral walls of the cavernous sinus, and the leukemic mass surrounding the carotid arteries (Figure 1b)

of the clivus and the cavernous sinus (Figure 1 a, b) and also patchy lesions in the calvaria. At the same time retroperitoneal lymph nodes and renal tumor masses were detected and the renal biopsy showed T-LBL tissue. Initially a CSF relapse was assumed, but CSF studies were normal.

The patient was in a critical condition, with severe uncontrollable headache, bilateral ptosis and diplopia. Due to the widespread intracranial lesions whole brain radiotherapy (12 x 2 Gy) was initiated. Despite a mild regression of tumor load especially in the calvaria and cavernous sinus in MRI symptoms progressed with severe headache and oculomotor deficits.

Due to the CN symptoms, which could be converged to the cavernous sinus, and also as an attempt to ameliorate the headache, the radiotherapy focus was changed targeting the base of the skull, clivus and cavernous sinus. The intended dose could not be completely given due to the deteriorating condition of the patient. An increase of the hepatosplenomegaly, lymph node mass and a pronounced hydronephrosis were described in abdomen CT.

Despite these efforts, and focal RT, CN symptoms progressed. The patient died due to sepsis. An autopsy was not possible.

Discussion

This observation is important for several reasons: firstly the focal recurrence of leukemia is rare, and even rarer without an associated leptomeningeal involvement. In this case, the patient had received prophylactic IT therapy, and during the time of the symptoms of the CSF, the CSF remained negative, which excluded meningeal spread.

The patient presented with severe neuropathic pain, which could hardly be controlled by drug treatment, and the focal RT was also intended as a symptomatic treatment for pain relief. Also sadly the severe oculomotor impairment and bilateral ptosis could not be improved and was a severe burden for the patient.

The cavernous sinus can be affected by several neoplastic processes at the base of the skull, mainly meningioma, rarely also as inflammatory processes [3]. In cancer the incidence of metastasis into the CS is appreciated to be less than 1 %. [4]. Retrograde infiltration via the cranial nerves has been observed [5].

Several cases of lymphoma presentations in the cavernous sinus have been reported, also bilaterally. Also myeloma can affect the CSF [6-10].

In leukemia the affection of the cavernous sinus has been reported in several cases [11, 12], often with involvement of adjacent structures. Also solid presentations of leukemia termed myeloid sarcoma or chloroma were described [13]. This case adds another observation to the bilateral involvement of the CN in leukemia's a solid deposit, and without CSF involvement presenting with intractable pain and visual symptoms.

Conclusion

This case has two important aspects: 1) local deposition of leukemia can occur in remission, and without CSF involvement. 2) The combination of oculomotor symptoms and therapy resistant pain suggest involvement of the cavernous sinus.

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