Hemepath Case 49: 9-Year-Old Boy

HISTORY

A 9-year-old boy is brought in by his mother. The child has been complaining of double vision and severe headaches since yesterday. He was diagnosed with acute lymphoblastic leukemia 3 years ago, but has almost completed his maintenance therapy without incident.

Right abducens nerve palsy and nuchal rigidity are demonstrated on physical examination. A lumbar puncture, CBC, and bone marrow aspirate and biopsy are quickly ordered. The peripheral blood smear and marrow findings are normal, showing no evidence of leukemia or any other lesion. The CSF is shown in the digitized slide below.

CBC

Hgb (g/L)	N
MCV	N
WBC	N
Plt	N

DESCRIPTION OF CSF

There is massive infiltration of the CSF by blast cells (see circles).

*** To see the slide annotations in Imagescope, click on VIEW, then ANNOTATIONS, and then on the "eye" icon adjacent to the word "Layers". In the “Layer Attributes” box, a brief description of the annotations is provided. You may also click on individual layer region (e.g. region 1) in the “Layer Regions” box to locate each annotation – this is especially helpful in identifying annotations when the slide is not zoomed in. ***

MORPHOLOGICAL DIAGNOSIS

Acute lymphoblastic leukemia, CNS relapse

DISCUSSION

This is a case of acute lymphoblastic leukemia (ALL) relapsing with CNS involvement only. Malignant cells infiltrate the meninges at the time of initial diagnosis via hematologic spread. Because of the blood-brain barrier, the CNS is a “protected” site: leukemic cells may therefore persist within the CNS even through chemotherapy. If these leukemic cells in the CSF manage to survive treatment, they may spontaneously

proliferate within the CNS even without relapsing in the blood or marrow. Subsequent elevation in intracranial pressure can give rise to headaches and papilledema.

CSF analysis will show an increased cell count and elevated protein levels, as well as the presence of malignant lymphoblasts. Treatment of patients with CNS leukemia (and prophylaxis against CNS leukemia) includes intrathecal chemotherapy and cranial irradiation.