Pseudotumor Cerebri in Patients with Acute Leukemia

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Background

- Pseudotumor cerebri (PTC), also known as idiopathic intracranial hypertension, is a clinical syndrome characterized by headache, vision changes, and papilledema on fundus examination.
- PTC can be primary, which is seen in obese women of childbearing age.
- PTC can also be secondary to Addison disease or certain medications such as all-trans retinoid acid (ATRA).

Materials and Methods

- We performed a single-center retrospective study of all acute leukemia (AL) patients with PTC at Memorial Hermann Hospital Texas Medical Center between January 1, 2013 and April 2022.
- Electronic medical records were used to collect baseline demographic data, clinical presentation, and outcomes of patients with PTC.

Results

- Among 127 patients with AL, 3 cases (2.3%) of PTC were identified.
- Table 1 shows clinical characteristics and presentation of patients with PTC.
- At our institution, we start empiric ATRA in all cases of suspected acute promyelocytic leukemia (APL). It is later discontinued if necessary after FISH results are back.

Results

- Patient 2 with acute myeloid leukemia (AML) had a history of PTC diagnosed 30 years previously that was treated with acetazolamide for 6 months; she had a recurrence of PTC 3 months after her diagnosis of AML.
- MRI brain in all 3 patients showed distension of the optic sheath, flattening of globe, cupping of papilla, and other features suggestive of PTC.
- Two patients (1 each with AML and acute lymphoid leukemia [ALL]) received intrathecal chemotherapy for CNS prophylaxis.

Results

- Cerebrospinal fluid cytology and flow cytometry showed no evidence of leukemia in any of the 3 patients.
- All 3 patients had improvement in headache with acetazolamide and lumbar puncture (LP) done for intrathecal chemotherapy.
- None of the 29 patients with APL developed PTC during treatment with ATRA.
- None of patients developed vision loss or other permanent neurological deficits.

Table 1 showing patient characteristics with PTC:

Patient	Age/ sex	BMI	Diagnosis	Timing of PTC diagnosis	Received empiric ATRA	Headache	Papilledema	MRI brain	LP-CSF opening pressure (cm of H_2O)
1	40/F	42	AML	Day 4 of induction chemotherapy	Yes, 3 doses	Yes	Yes	Abnormal	35
2	52/F	44	AML	3 months after diagnosis of AML	Yes, 3 doses	Yes	Yes	Abnormal	NA
3	38/F	42	Ph+ ALL	3 months after diagnosis of ALL	No	Yes	Yes	Abnormal	30

Discussion

- PTC is a rare event in patients with AL.
- Diagnosis of PTC is based on findings of papilledema, normal neurological exam except for cranial nerve deficits, and neuroimaging showing an absence of mass, lesion, or hydrocephalus.
- Diagnostic LP to measure opening pressure is challenging in AL patients due to severe thrombocytopenia and coagulopathy.
- Patients with “probable” PTC who cannot safely undergo LP should have treatment initiated promptly.

Conclusion

- PTC is a rare diagnosis and can complicate the clinical course of AL patients.
- In addition to ATRA-induced PTC, clinicians should also consider the possibility of primary PTC, which is seen in morbidly obese women of childbearing age.
- Patients with a prior history of PTC may experience recurrence during AL therapy.

References

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