Acute Promyelocytic Leukemia: A Case of Pseudotumor Cerebri Syndrome During Treatment Consolidation

Alexandra Ghiaur MD[1], Bogdan Ionescu MD[1], Cristina Constantin MD[1], Didona Vasilache MD[1], Mihaela Dragomir MD[1], Daniel Coriu PhD, MD[1,2]

[1]Fundeni Clinical Institute, Bucharest, Romania. [2]Carol Davila University of Medicine and Pharmacy, Bucharest, Romania

Background

Pseudotumor cerebri, also known as idiopathic intracranial hypertension, describes a neurologic syndrome characterized by headache, visual changes, papilledema without cerebral mass lesion, venous sinus thromboses, or obstructive hydrocephalus. Risk factors for the development of pseudotumor cerebri are obesity, women in the childbearing years, and hypervitaminosis A.

The modified Dandy criteria are used for pseudotumor cerebri syndrome(PTCS) diagnosis. This complication has a high incidence in the pediatric population, particularly within 2–3 weeks of induction treatment for APL.The reported incidence of “probable” PTCS is 1.7% in APL patients treated with ATRA (all-trans-retinoic acid)

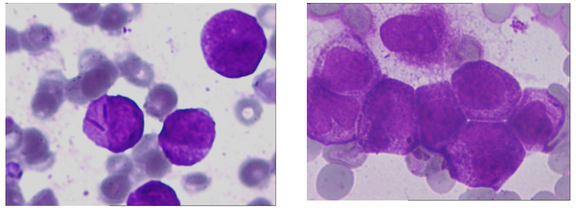


Figure 1. Peripheral blood smears showing the presence of atypical promyelocytes wth numerous Intracytoplasmatic Auer rods (May-Grumwald-Giemsa stain, original magnification x100)

Case Presentation

We report here the case of a 20-year-old woman, obese with a BMI of 34.5 kg/m^2, diagnosed with low-/intermediate-risk APL and treated with ATRA and ATO as induction treatment. After induction, she achieved MRD-negative remission.

During the first 2 weeks of the first cycle of consolidation—ATRA and ATO—the patient developed progressive diplopia and headaches without response to standard analgesic therapy. Additionally, she complained of nausea and vomiting. The ophthalmological exam with fundus examination revealed bilateral papilledema. The neurological exam was normal, except of bilateral sixth cranial nerve palsy.There were no abnormal findings in the head CT scan orMRI of the brain. Thus, a diagnosis of “probable” pseudotumor cerebri was established

Intervention and Clinical Course

ATRA was discontinued and she received acetazolamide and dexamethasone with resolution of neurological symptoms within the next 7 days.

Upon the next consolidation ATRA and ATO, the headaches resumed. The patient received all the ATRA consolidation cures as scheduled but with a dose reduction to 80% of the initial dose and in association with acetazolamide.

She completed consolidation treatment and continues to be in molecular complete remission.

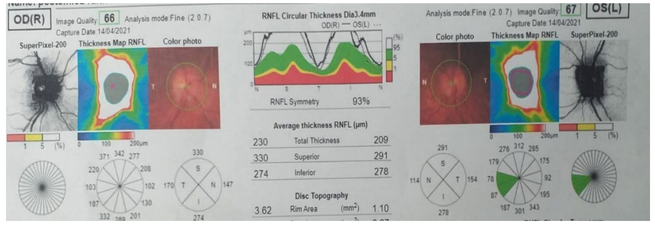


Figure 2A. Initial diagnosis of PTCS . Color fundus photos from initial diagnosis showed bilateral papilledema with friscen grade 2 on right eye and grade 3 on left eye. Optical coherence of tomography (OCT) of optic disc revealed increased average retinal nerve fiber layer (RNFL) thickness of both eyes.

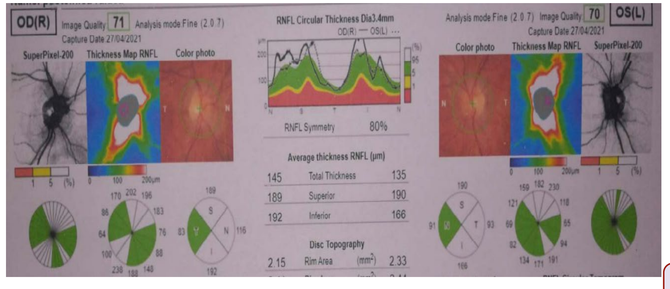


Figure 2B. Assessment after 10 days of treatment Fundoscopy and optical coherence tomography (OCT) with RNFL thickness map was used to monitor the response to treatment.. Fundoscopy revealed friscen grade 1 of bilateral papilledema. The averange RNFL was significantly lower in both eyes.

Summary

This case report highlights the possibility of pseudotumor cerebri development in young adult patients with APL during consolidation therapy. The neurological symptoms were reversible, and the patient received all the ATRA consolidation. It is important to exclude other causes of intracranial hypertension including thromboses, infections, and disease involvement

Discussion

Through a literature review PTCS was described following ATRA administration, as well during the concomitent use of ATRA and azole drugs. Early management of this rare complication is essential to prevent blindness.

There is no standard recommandation, therefore the management strategies consist of discontinuation of ATRA/ATO, acetazolamide, diuretics, therapeutic lumbar punctures, dexamethasone.

References

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