Author's response to reviews

Title: intracranial extramedullary hematopoiesis in Beta Thalassemia; case report

Authors:

Homayoun Tabesh (homtabesh@yahoo.com)
Ahmad Shekarchizadeh (shekarchizadeh@med.mui.ac.ir)
Parvin Mahzouni (Mahzouni@med.mui.ac.ir)
Mojgan Mokhtari (mokhtari@med.mui.ac.ir)
Saeid Abrishamkar (abrishamkar@med.mui.ac.ir)
Salman Abbasi Fard (dr_abbasi_s@yahoo.com)

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Author's response to reviews: see over
Dear Reviewers

With Due Respect, first of all we should appreciate for considering our manuscript.

Reply to Reviewer 1:

- To explain why the patient would have loss of vision in both eyes, we should say that the visual loss in our case may be due to secondary effects of increased ICP and/or mass lesion upon adjacent structures of visual pathways. Moreover, intracranial extramedullary hematopoiesis arising from the base of the cranium near the sphenoid and ethmoid bones, may extend into the intracranial cavity, and compress the visual pathways with resultant progressive visual failure.

- The patient’s identifiable information is removed from the MRI images.

Reply to Reviewer 2:

- To answer the questions No. 1 to 3 we should explain that, as far as our patient was a known case of thalassemia we did hematologic consultation and according to his note the EMH was one of the differential diagnoses. But in our case the findings were not sufficient to confirm the diagnoses of EMH and therefore the subsequent treatment with radiation therapy (by considering this fact that the extra medullary hematopoietic masses are highly sensitive to radiotherapy). Moreover, other entities should be considered in the differential diagnosis including lymphoma, neurosarcoïdosis and etc. Additionally, because of the signs and symptoms of increased intracranial pressure secondary to direct mass effect in our case, we decided to perform an emergent neurosurgical intervention.

- According to hematologist consultation, Laboratory investigations were; hemoglobin 8 g/dL, WBC count: 13.2x10^9/l, platelet count: 265x10^9/l, mean corpuscular volume: 68.9 fl. Peripheral smear showed anisopoikilocytosis, tear drop cells, polychromasia, nucleated RBCs and target cells. Hemoglobin electrophoresis showed 96.7% HbF and 3.3% HbA2. HbA was absent. In his family history his parents were both carrier (minor thalassemia). The patient did not consume any kind of medicine and his past medical history is written in our manuscript.

- Postoperatively, patient was followed for 4 months. He showed rapid recovery of signs and symptoms of increased intracranial pressure but incomplete recovery of visual lost i.e. Visual acuity was improved to detect hand motion. During this period patient was referred to hematologist again and radiotherapist for continuation of his treatment.
Just as mentioned in the last paragraph of case presentation, patient received standard treatment of EMH by hematologist.

Reply to Reviewer 3:

- The patient’s identifiable information is removed from the MRI images.
- With special thanks for your suggestion, we added both articles to our references and we addressed them.

Corresponding Author:

Salman Abbasi Fard, Neurosurgery department, Al-Zahra Hospital, Isfahan, Iran.
Tel: +98-912-3931633
Fax: +98-311-6685555
Email: Dr_abbasi_s@yahoo.com