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MAGNETIC RESONANCE IMAGING OF INTRACRANIAL EXTRAMEDULLARY HAEMATOPOIESIS IN β-THALASSEMIA: A CASE REPORT

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ABSTRACT

We report the Magnetic Resonance Imaging of intracranial extramedullaryhematopoiesis in 21 years old β-thalassemic patient who presented to the Neurology Department of Civil Hospital Karachi with seizures. The MR exam revealed multiple intracranialextra-axial masses;isointense on T1W sequence and hypointenseon T1W, T2W, FLAIR, DWI sequence and ADC (b value= 1000 sec/mm2) with avid post contrast enhancement. Plain CT of the Head documented the presence of multiple hyperdenseextra-axial masses with mean value of 45 HU. Extramedullary hematopoiesis is the formation and development of blood cells in tissues outside of the medulla of the bone. The most common sites of involvement are the liver, spleen, and lymph nodes.Intracranial extra medullary hematopoiesis is a relatively rare manifestation of Thalassemia. There is paucity of literature for MR findings regarding intracranial EMH. It is essential to distinguish it from other central nervous system tumors, because treatment and prognosis are entirely different.

Key words: β-thalassemia;EMH; MRI; DWI; ADC; intracranial extramedullary hematopoiesis

INTRODUCTION

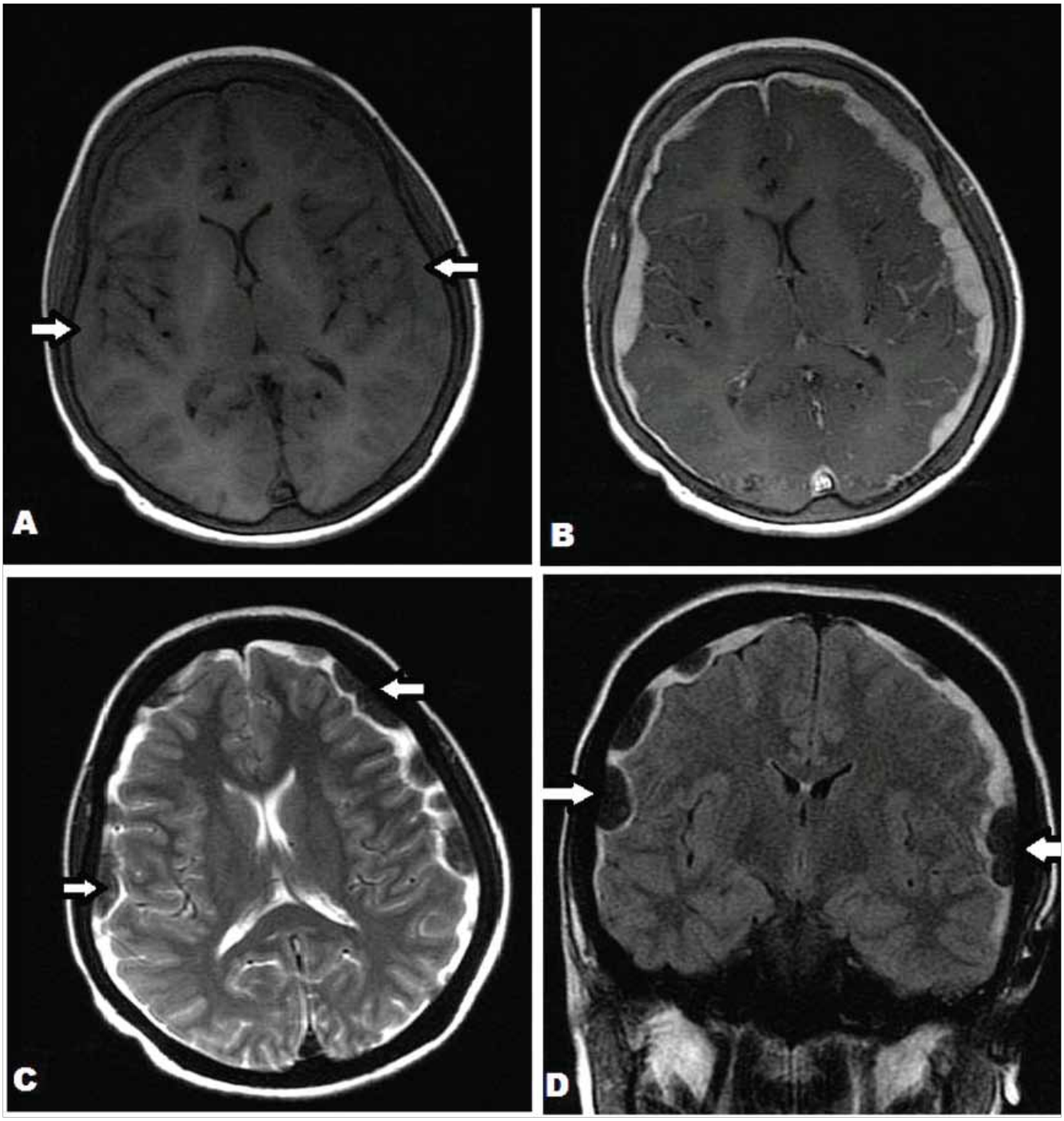
Thalassemia is among the most common genetic disorder worldwide. Health experts claim that approximately 5,000 children are diagnosed with thalassemia-major, every year in Pakistan. Beta-thalassemia is a hemolytic disease characterized by a gene defect in the production of beta globin chain from hemoglobin1, which causes ineffective erythropoiesis and peripheral destruction of red blood cells. Extramedullary hematopoiesis occurs in approximately 15% of cases of thalassemia2.It is a process in which the body attempts to maintain erythrogenesis by proliferation of pluripotent stem cells outside the bone marrow due to less production of blood cells in order to meet the body's demand to compensate for anemia. Apart from thalassemia, it is also observed insickle cell anemia, leukemia and in myeloproliferative disorders such as myelofibrosis, polycythemia rubravera3. EMH occurs most often in the spleen and liver and occasionally in the lymph nodes. Less common organs include the pleura, lungs, gastrointestinal tract, breast, skin, kidneys, and adrenal glands4. Intracranial extra medullary hematopoiesis is a relatively rare manifestation of Thalassemia. It can possibly involve the cranial dura, falx, cerebral parenchyma, optic nerve sheath, choroid plexus and the epidural space of the spinal canal. Usually patients are clinically silent, headache, seizures, visual disturbance, papilledema, motor and sensory impairment, and even coma may develop5. Magnetic resonance imaging (MRI) remains the modality of choice for its diagnosis for its diagnosis6; being a noninvasive technique.We present a rare case of intracranial EMH in 21 years old thalassemic patient.

CASE SUMMARY

A 21-year-old Asian male, with known beta thalassemia, presented to Civil Hospital with complaints of low grade fever and generalized epileptiformtonic clonic seizures during the last three weeks, with recent onset headache, nausea, vomiting and blurring of vision for last 3 days.His personal history was significant for irregular blood transfusions. He was also known case of bronze diabetes. There was documented history of weight loss for last one month. The family history revealed that his parents werecarriers (minor thalassemia) and his two younger sisters were also known thalassemic major.He had pallor of the skin with a cachectic appearance and a prominent skull. The patient has hyper pigmented dermal plaques over the arms and abdomen.Neurological examination wasunremarkable except for bilateral mild papilledema demonstrated atfundoscopy. On admission, laboratory investigations showed hemoglobin 8 g/dl, white blood

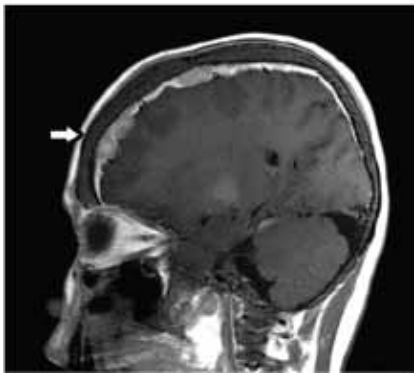
cell count 5.33 × 109/L, platelet count 118 × 109/L and mean corpuscular volume 77.8fl. Ultrasound exam revealed gross hepato-splenomegaly with diffuse increased echogenicity of liver with coarsening of liver parenchyma and atrophic pancreas.Magnetic resonance imaging of the brain was performed on GE Health Care SignaHDxt1.5 Tesla Scanner,using the head coilwith slice thickness of 4mm with interslice width of 1mm. Multiplanar, multisequential images in T1W, T2W fast spin echo sequence, FLAIR and single shot echo planner DWI sequence at a b-value of 1000 sec/mm2, along with post contrast T1W sequence were obtained. MRIrevealed multiple well defined intracranial extra-axial masses

Figure 1: Axial T1- (A), post contrast axial T1 weighted- (B), axial T2 weighted Fast Spin Echo - (C) and FLAIR coronal (D) images demonstrate iso-intense T1 signal, low T2 and FLAIR signal intradural masses. The lesions shows enhancement in post contrast T1-weighted image (D) –[arrows in Fig A, C & D].



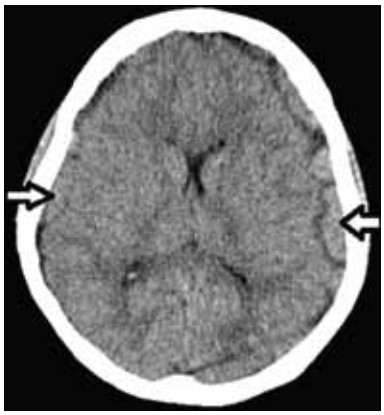
(Figure 1) over the cerebral convexities bilaterally which were isointense to gray matter on T1-weighted sequence and hypointense on T2-weighted, FLAIR sequence, Diffusion Weighted imaging and apparent diffusion coefficient (ADC) map with avid homogenous enhancement on post contrast T1-weighted sequence.The maximum width of these masses measures 20mm in transverse dimensions.No adjacent parenchymal edema was seen. It was associated withdiffuse pachymeningeal thickening and enhancement.There was widening of diploic space of the calvarium which demonstrated hypointense signals on all sequences due to replacement of fatty marrow with hematopoietic marrow as a compensatory mechanism (Figure 2).

Figure 2: Para-Sagittal post contrast T1 weighted image shows widening of the diploic space [arrows] with hypointense marrow signals.



The patient underwent plain brain CT of head (Fig.3) performed with 16 slice Toshib

Figure 3: Plain CT Head shows multiple iso to hyperdenseintradural masses [arrows]
piral CT [scanning parameters of 120 KV, 150mA and 5mm slice thickness] which showed multiple iso to hyperdenseintradural masses with mean value of 45 HU.



DISCUSSION

Our case describes 21 years old thalassemic patient with history of irregular blood transfusion and now presented with GTC seizures for short duration of time. MR Brain with epilepsy protocol was done to identify the epileptogenic focus. His MRI demonstated multiple extra-axial T1 isointense and in particularly T2,FLAIR, DWI hypointense masses with avid post contrast enhancement. The T2 blackout phenomenon was noted in our case which leads to hypointensity on DWI and T2W images. BivekKarki et al1 in his case report for similar signal intensity lesion13-year-old beta-thalassemic patient with left sided paresis, initially proposed the differential diagnosis of multiple meningioma, dural metastasis, lymphoma and granuloma. It was emphasized that meningioma, lymphoma and metastasis demonstrate T2 hyperintense signals and meningioma has a tendency to occur in middle aged female patients; therefore excluded. Granuloma occurs in known cases of CNS infective orinflammatory disorders – not compatible to our case. T2-hypointensity of the lesions on MRI is characteristic for deoxyHb, intracellular metHb and hemosiderin7. Hemoglobin being a paramagnetic substance, evolves in time from oxyHb, then deoxyHb to metHb converting into ferritin and finally to hemosiderin. Higher concentrations of hemosiderin can cause a strikingly low signal on T2W sequence8.The cause of T2 blackout demonstrated in our case is predominantly due to susceptibility effect of these paramagnetic substances9. The signal characteristics of the enhancing lesions on MRI signify the presence of cells containing blood/iron products. Extramedullary hematopoiesis (EMH) refers to formation of blood cells occurring outside the medulla of the bone, secondary to insufficient bone marrow function10.Diagnosis of EMH is based on clinical features, imaging modalities and histopathology. HomayounTabeshand his colleagues2reported a case of 34-year-old thalassemic Caucasian man with the complaints of chronic headache whose MRI of his brain revealed a hyperintense lesion on T1W images which was signal void on T2W images in the left trigone area with vivid post contrast enhancement. Koch et al5reported T1W isointense and T2W markedly hypointenseintradural mass at posterior interhemisphericfissue showing diffuse enhancement in known hematopoietic disorder with symptoms of weakness, numbness, and hyperreflexia. The signal intensity of these intracranial masses in the above mentioned studies is compatible to our case. All studies including this case provided the evidence that MRI has evolved as the modality of choice in the diagnosis of intracranial EMH. Patients

can directly undergo radiotherapy without suffering from morbidity of surgical biopsy as hematopoietic elements are intensely radiosensitive11. Surgical intervention should be reserved only for uncertain cases6. Therare presentation of seizures for intracranial EMH along with multiplicity of the intracraniallesions was unique to our case. This study emphasizes the fact that characteristic T2W, FLAIR and DWI hypointensity of the enhancing masses on sequence is due to magnetic susceptibility effect, caused by accumulationof blood products within tissue, further confirmed by demonstration of high attention soft tissue masses on CT.

CONCLUSION

This case along with literature review highlights the importance of Magnetic Resonance Imaging as the modality of choice for the diagnosis of intracranial extramedullary hematopoiesis - a rare manifestation in known hematopoietic/ myeloproliferative disorder. It is essentialfor the radiologist to become familiar with the MR features of intracranial EMH for correct diagnosis treatment and prognosis is entirely different from other CNS tumors.

REFERENCES

1. Marengo-Rowe AJ. The thalassemias and related disorders. InBaylor University Medical Center. Proceedings 2007 Jan 1 (Vol. 20, No. 1, p. 27). Baylor University Medical Center.
2. Tabesh H, Shekarchizadeh A, Mahzouni P, Mokhtari M, Abrishamkar S, Fard SA. An intracranial extramedullary hematopoiesis in a 34-year-old man with beta thalassemia: a case report. Journal of medical case reports. 2011 Dec 19;5(1):1.
3. Haidar S, Ortiz-Neira C, Shroff M, Gilday D, Blaser S. Intracranial involvement in extramedullary hematopoiesis: case report and review of the literature. Pediatric radiology. 2005 Jun 1;35(6) :630-4.
4. Koch BL, Bisset 3rd GS, Bisset RR, Zimmer MB. Intracranial extramedullary hematopoiesis: MR findings with pathologic correlation. AJR. American journal of roentgenology. 1994 Jun;162(6): 1419-20.
5. Tayari N, Ahrar MH, Jafarpishe MS. Case report of the extramedullary hematopoiesis presented as a hypervascular intracranial mass. Advanced biomedical research. 2013;2.
6. Karki B, Xu YK, Tamrakar K, Wu YK. Intracranial extramedullary hematopoiesis in beta-thalassemia. Korean journal of radiology. 2012 Apr 1;13(2):240-3.
7. Zimny A, Neska-Matuszewska M, Bladowska J, Sąsiadek MJ. Intracranial Lesions with Low Signal Intensity on T2-weighted MR Images–Review of Pathologies. Polish Journal of Radiology. 2015;80:40.
8. Jason K Lee, D William, Boswell. Extramedullary Hematopoiesis. ApplRadiol. Medscape. 2005 Oct;34(10):44-47.
9. Hiwatashi A, Kinoshita T, Moritani T, Wang HZ, Shrier DA, Numaguchi Y, Ekholm SE, Westesson PL. Hypointensity on diffusion-weighted MRI of the brain related to T2 shortening and susceptibility effects. American Journal of Roentgenology. 2003 Dec;181(6):1705-9.
10. An J, Weng Y, He J, Li Y, Huang S, Cai S, Zhang J. Intrathoracicextramedullary hematopoiesis presenting as tumor simulating lesions of the mediastinum in α-thalassemia: A case report. Oncology letters. 2015 Oct 1;10(4):1993-6.
11. Lee CM, Salzman KL, Blumenthal DT, Gaffney DK. Intracranial extramedullary hematopoiesis: brief review of response to radiation therapy. American journal of hematology. 2005 Feb 1;78(2):151-2.

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Author's contribution:
MahnoorHafeez; Study concept and design, protocol writing, data collection, data analysis, manuscript writing, manuscript review
Ateeque Ahmed Khan; data collection, data analysis, manuscript writing, manuscript review