Case Report

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Extramedullary Hematopoiesis Presenting as Progressive Headache: A Case Report and Review of Literature

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Citation Babaee MR, Mohseni I, Mohammadi-Vajari MA, Tefagh Gh, Rakhshankhah N, Motevalli M. Extramedullary Hematopoiesis Presenting as Progressive Headache: A Case Report and Review of Literature. Case Reports in Clinical Practice. 2021; 6(4):124-136.

Running Title: Extramedullary Hematopoiesis Presenting as Progressive Headache

Article info:
Received: 01 July 2021
Revised: 11 July 2021
Accepted: 03 August 2021

Keywords:
Hematopoiesis; Extramedullary cell differentiation; Thalassemia

ABSTRACT

Extramedullary Hematopoiesis (EMH) is defined as the production of blood cells in organs other than bone marrow. Intracranial EMH is a rare condition. In this article, we presented a case of intracranial EMH presenting as progressive headache. Our patient was a 33-year-old man with thalassemia presenting with acute progressive flaccid quadriplegia, severe progressive headache, and decreased level of consciousness. His imaging studies showed evidence of intracranial and presacral EMH. Most asymptomatic intracranial EMH can present as a variety of symptoms, including progressive headache; therefore, the differential diagnosis should be kept in mind when evaluating a patient with a relevant underlying medical condition.

Introduction

Hematopoiesis is defined as the formation of blood cells from hematopoietic stem cells, which reside in the bone marrow. Extramedullary Hematopoiesis (EMH) is a condition, in which various organs in the body (other than the bone marrow) play a role in the hematopoiesis. The usual sites of EMH are known to be the spleen and liver probably due to their role in hematopoiesis during embryonic and fetal development [1]. Other less common sites of EMH have been reported to be the paraspinal region, abdominal viscera, thymus, pleura, and also intracranial structures [2].

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Intracranial hematopoiesis is a rare condition. Thalassemia (50%) and myelofibrosis (31%) are two common causes of intracranial involvement by EMH [3]. Frequent sites of intracranial EMH are the dura mater, the brain parenchyma, the sheath of the optic nerve, and the bony skull, respectively [4].

Intracranial EMH is usually found in asymptomatic patients or patients with symptoms unrelated to intracranial EMH; however, symptoms, such as headache, confusion, gait disturbances, and subdural hematoma have been linked to intracranial EMH [5]. Here, we presented a rare case of simultaneous intracranial and paraspinal EMH presenting with progressive headache and flaccid quadriplegia.

Method of Review

A literature search was done to find relevant case report articles in English in PubMed® and also available abstracts of the related new congresses. Used keywords included intracranial extramedullary hematopoiesis (intracranial [All Fields] AND ("extramedullary haematopoiesis" [All Fields] OR "hematopoiesis, extramedullary" [MeSH Terms] OR "hematopoiesis" [All Fields] AND "extramedullary" [All Fields] OR "extramedullary hematopoiesis" [All Fields] OR "extramedullary" [All Fields] AND "hematopoiesis" [All Fields]) AND Case Reports [ptyp]. Also, another search of bibliographies of the results was done to find articles that were not included in the primary search.

In the primary review, 39 articles were found And after primary evaluation, 31 articles were included in the study. Other articles did not fulfill the critical appraisal checklist for case reports. Age, sex, presenting symptom of the patients the diagnostic modality used, and the features of the lesions on Computed Tomography (CT) scan, Magnetic Resonance Imaging (MRI), other imaging modalities used to evaluate the lesion, as well as the treatment and outcome of each case are summarized in Table 1.

Case Presentation

Our patient was a 33-year-old man diagnosed with thalassemia major from childhood presented to the emergency ward with the chief complaint of acute progressive flaccid quadriplegia, severe headache, and decreased level of consciousness. On admission, he had a hemoglobin level of 6.6 g/dl. He had not received his transfusions regularly due to poor compliance. An emergency brain CT scan was ordered to investigate the cause of his decreased level of consciousness (Figure 1).

Multiple mass-like lesions with a solid and partially dense appearance were seen on both sides of the interhemispheric falx in an extra-axial and dural-based position along with similar lesions adjacent to the convexity of the right frontal lobe. Considering the patient's past medical history, this was suggestive of EMH. However, the lesions on the brain CT scan could not explain his flaccid quadriplegia. To investigate the possible cause of the quadriplegia, a spiral abdominopelvic CT scan and a cervicothoracic spinal MRI were done. In the CT scan, diffuse bony expansion and coarse trabeculation suggestive of bone marrow hyperplasia were seen in the ribs, vertebrae, and pelvic bone. A solid mass-like lesion with homogenous enhancement with a size of 95×59×78 mm was seen in the presacral space extending to the anterior epidural space (Figure 2).

The compression effect of this mass had caused sacral spinal canal stenosis. Considering the patient's background, this was also suggestive of EMH. In the cervicothoracic spine MRI (Figure 3), diffuse low signal bone marrow as well as bony hypertrophy were seen in the thoracic vertebrae. These changes had caused thoracic cord compression, which was apparent as patchy high signal areas on the T2-weighted MRI. Lesions with low signal in the posterior thoracic epidural space on T1- and T2- weighted sequences were suggestive of EMH. It seemed that the compression effect of the lesions seen on the abdominopelvic CT scan and the cervicothoracic spine MRI, was the cause of his quadriplegia; therefore, a series of corticosteroid pulses were started for him.

Unfortunately, despite the admission of corticosteroid pulses, the patient's clinical course did not show any improvement. His level of consciousness did not improve and he also developed respiratory distress leading to intubation. Unfortunately, he developed hyperkalemia and metabolic acidosis in the course of his disease and dialysis was not feasible due to his increased International Normalized Ratio (INR) and low platelet count. A Peripheral Blood Smear (PBS) was drawn to rule out Disseminated Intravascular Coagulation (DIC) as the cause of the disturbance in his coagulation studies, in which no sign of schistocytes was detected. On the fifth day of his admission, he proceeded to hypotension and asystole, and resuscitation was not successful.

Discussion

EMH is the production of blood corpuscles in body parts other than bone marrow. This condition has been described to occur physiologically in the course of fetal development and also as a part of the immune response

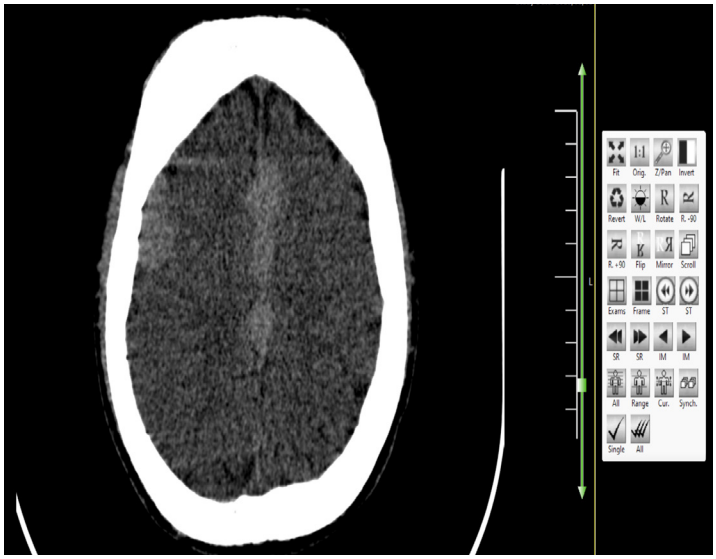

Figure 1. Non-contrast brain Computed Tomography (CT) scan

Multiple solid partially hyperdense mass-like lesions with the extra-axial dural-based position on both sides of the interhemispheric falx and convexity of the right frontal lobe are observed.

during some infections [6]. Pathologic EMH usually occurs as a response to either ineffective hematopoiesis seen in conditions, such as myelofibrosis, or hyperactive bone marrow response in hemolytic anemias, such as thalassemia and sickle cell anemia [6, 7]. It has been proposed that the increased need for blood cells leads to the proliferation of pluripotent stem cells, which reside in mesenchymal organs [8].

Although compression of the spinal cord by EMH is a familiar condition [9], intracranial presentation is particularly uncommon and there have been few published cases of this situation. Intracranial EMH has been reported to occur in the diploic space [10, 11], epidural space [5, 10], cerebral parenchyma [12], and the falx cerebri [13].

The necessary step in the successful diagnosis of intracranial EMH is considering the patient's underlying

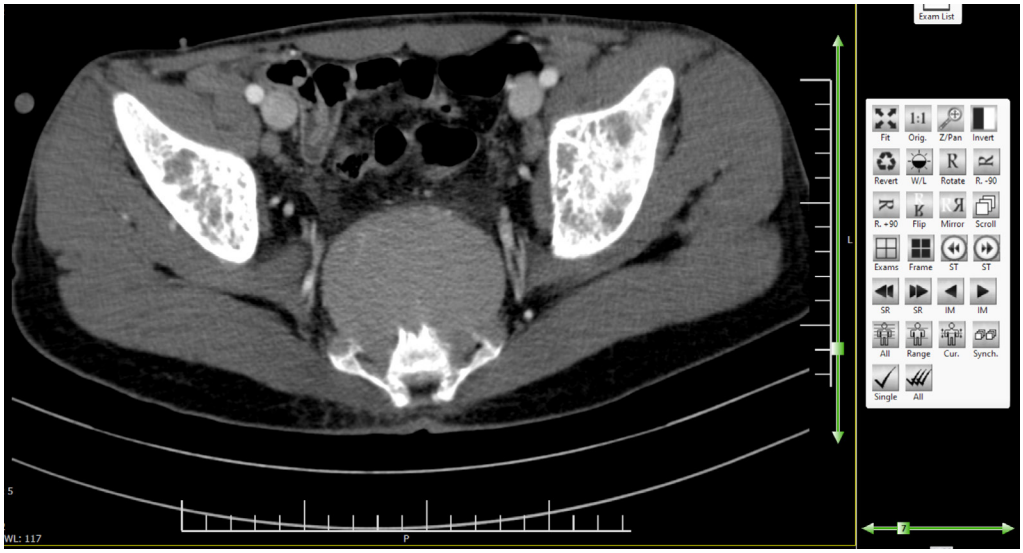

Figure 2. Contrast-enhanced abdominopelvic Computed Tomography (CT) scan

There is a solid mass-like lesion with homogenous enhancement in the presacral space extending to the anterior epidural space. There is also evidence of bone expansion and coarse trabeculation of iliac bones.

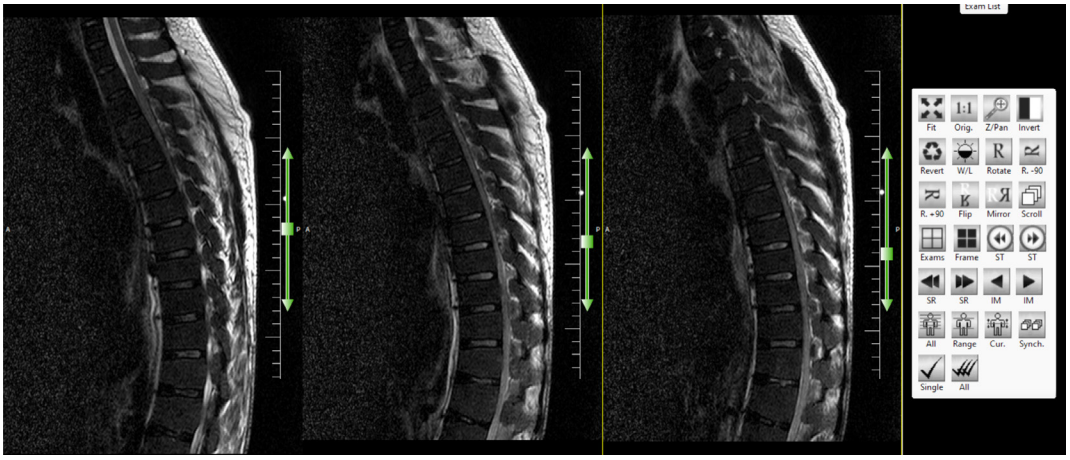

Figure 3. Cervicothoracic spine magnetic resonance imaging (MRI)

Diffuse low-signal bone marrow and bony expansion are seen in the thoracic vertebrae. These changes had caused thoracic cord compression, which was apparent as patchy high-signal areas on the T2-weighted MRI.

disease, symptoms, and imaging findings (Table 1). Intracranial EMH has been mostly seen in patients with a history of thalassemia major, intermedia, and myelofibrosis. Other less common underlying diseases are polycythemia vera, myelodysplastic syndrome, multiple myeloma, and Central Nervous System (CNS) lymphoma (Table 1).

One of the most common encountered symptoms in symptomatic patients with intracranial EMH is headaches associated with other signs of increased intracranial pressure. Other presentations are those affecting the cerebral cortex, such as seizures, loss of consciousness, and upper motor paralysis (Table 1). Our patient also suffered from severe progressive headache, which is the most common symptom reported in the literature.

Different imaging modalities are used for the diagnosis of EMH. The CT appearance of EMH is a heterogeneous lobulated soft tissue density mass. MRI is the modality of choice for the diagnosis of intracranial EMH. Intracranial EMH is defined as unique or multiple iso-intense or hyper-intense extra-axial appended masses to the meninges, which represent homogeneous enhancement after contrast administration. These masses often appear as lobular and well-defined masses with intermediate signal intensity on T1-weighted and low signal intensity on T2-weighted images. Intracranial EMH may represent notable enhancement following gadolinium injection. Also, a high-signal intensity rim is seen, which represents fatty tissue on both T1- and T2-weighted images [3, 14, 15]. Due to our patient's both critical situation and renal failure, performing a contrast-enhanced MRI was not feasible. However, in his non-contrast imaging, compatible findings with the literature were found (Figures 1, 2, 3, and 4 and Table 1).

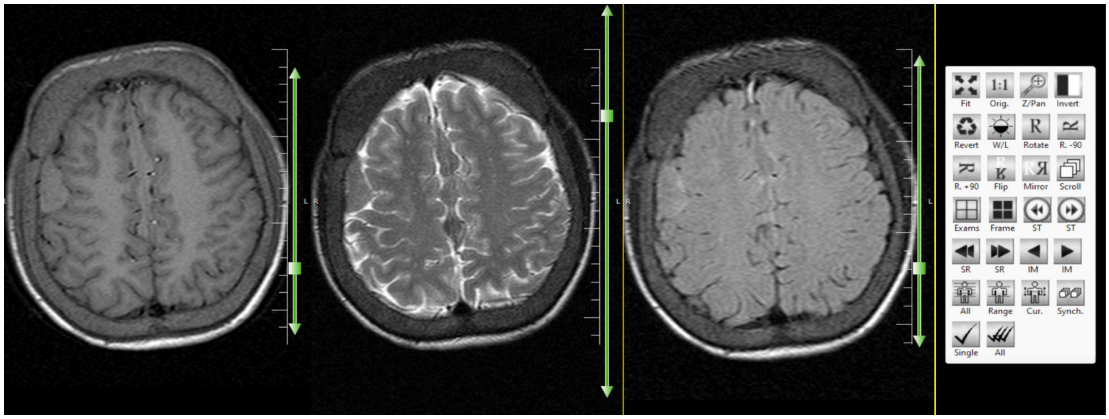

Figure 4. Non-contrast brain Magnetic Resonance Imaging (MRI)

There are some epidural mass-like lesions in the right frontoparietal region and a subdural mass-like lesion in the interhemispheric fissure, which are isointense in T1-/T2-weighted and Fluid-Attenuated Inversion Recovery (FLAIR) images. Bone expansion of the bony skull is seen.

Table 1. Case reports of the intracranial Extramedullary Hematopoiesis (EMH)

PMID	Author, Year	Age/Sex	Presenting Symptoms	Underlying Disease	CT Features	MRI Features	Treatment	Outcome
29368108	Merchant et al. (2018) [10]	18/m	Headache crisis for 2 years Vomiting episodes	Beta-thalassemia major with no regular transfusions	NM	Frontal and parietal bilateral even, epidural soft tissue density Isointense on T1-weighted images and hypointense on T2-weighted images Effacement of sulci and lateral ventricles Brainstem pushing by herniation of the tonsils and tentorium	Blood transfusion Hydroxyurea	Symptoms improved
		17/m	Headaches since 6 months	Beta-thalassemia major	NM	Diffuse hypointensity of calvarium on T1- and T2--weighted images Indentation of the right parieto-occipital lobes, increasing and bulging of the right parietal calvarium	Blood transfusion Hydroxyurea	Symptoms improved
28681787	Reddy (2017) [16]	40/m	Headache and diplopia for 2 weeks	Myelofibrosis	CT only indicated the lytic nature of subcentimetric enhancing lesions of bone	T1-weighted images hypointense parafalcine and paratentorial lobulated soft tissue lesions, which showed gradient blooming on gradient recalled echo (GRE) T2-weighted images and homogenous enhancement after intravenous injection of gadopentetate dimeglumine No perilesional edema The calvarium was thickened with diploic space widening and showed multiple subcentimetric well-circumscribed enhancing lesions	NM	NM
28603913	Ruiz-Cordero et al. (2015) [17]	65/m	Headaches and right facial numbness	Primary myelofibrosis	NA	Focal leptomeningeal involvement Abnormalities were seen in acute leukemia *CSF and bone marrow flow cytometric analysis indicated leukemic leptomeningeal involvement	NA	NA
25812022	Mehta et al. (2015) [18]	67/f	Falling down with confusion (GCS=12) drowsiness	Beta-thalassemia minor and myelodysplastic syndrome	Left SDH	NM	Craniotomy biopsy*: Analysis of dura and membrane specimens showed EMH Radiotherapy	Death

PMID	Author, Year	Age/Sex	Presenting Symptoms	Underlying Disease	CT Features	MRI Features	Treatment	Outcome
23977662	Tayari et al. (2013) [11]	34/m	Progressive headache and epilepsy Nausea, vomiting and loss of vision	Beta-thalassemia intermediate	Enhanced hyperdense Left paraventricular mass with peripheral edema External and internal skull tables and diploic space thickening	Hyperintense lesions on T1-weighted images and hypointense on T2-weighted images T1-weighted images with contrast showed intense and homogeneous enhancement	NM	NM
23621819	Singer and Quencer (2014) [5]	54/m	Headaches last 10 days	Idiopathic myelofibrosis	Nodular epidural hyperdensity in the cerebral falx, cerebellar tentorium, and cerebral dura	Nonenhanced T1-weighted images: nodular epidural isointense mass was seen in both frontoparietal lobes, cerebral falx, and cerebellar tentorium Enhancement was seen in nodular extra-axial masses T2-weighted and FLAIR images: The absence of edema in the underlying brain parenchyma SWI: susceptibility was seen in mass due to iron deposition	Radiotherapy	Symptoms improved but then, progressive cognitive decline and worsening anemia happened due to radiotherapy
23610149	Palma et al. (2013) [19]	77/f	Obtundation, drowsiness, and disorientation for 15 days	Multiple myeloma	Multiple extra-axial hyperdense foci without bone destruction	NM	Platelet transfusion	Deth *Autopsy confirmed intracranial extramedullary hematopoiesis
22438693	Karki et al. (2013) [8]	13/m	Gait disturbance for 1 year	Beta-thalassemia with intermittent transfusion	High attenuating mass in the right temporoparietal lobe, which caused compression on ventricle and midline shifting Diploic space was thickened	Slightly hyperintense lesion on T1-weighted and hypointense on T2-weighted and FLAIR images with no perilesional edema DWI: homogeneous hypointensity consistent with diffusion coeffcient (ADC) Contrast-enhanced images: Apparent enhancing of the dura and intermediate enhancing of mass was seen	Surgery	NM

PMID	Author, Year	Age/Sex	Presenting Symptoms	Underlying Disease	CT Features	MRI Features	Treatment	Outcome
22220514	Eskazan/2012 [20]	30/m	Progressive headache	Beta thalassemia major	NM	Interhemispheric fissure mass measuring 4 cm×5 cm×11.5 cm	Hypertransfusion therapy hydroxyurea at a dose of 1000 mg/day	Regression of the interhemispheric EH mass after 6 months Follow-up by control cranial MRI
21854477	Jiang/2011 [21]	37/f	Worsening visual problems for 8 months	NM Lab data*: WBC:15.9* /L Plt: 92*/L RBC : 2.59*/L; Hb : 88 g/L MCV: 106 fl MCH: 35 pg MCHC : 330 g/L RDW:29%	NM	Bilateral frontotemporal and falx cerebral masses intermediate intensity on T1-weighted images and low-signal intensity on T2-weighted images with homogeneous enhancement but no perilesional edema Hypointensity on T1-weighted images and T2-weighted images with heterogeneous enhancement was observed in clivus	Surgery radiotherapy	NM
21175566	Eskazan/2011 [20]	30/m	Progressive headache	Beta-thalassemia major with intermittent transfusion	NM	A mass in the interhemispheric fissure	Blood transfusions Hydroxyurea	Symptoms improved
17762725	Zona/2007 [23]	58/m	Sudden onset of generalized seizures Left hemiparesis	Idiopathic myelofibrosis	Heterogonous right frontal lesion with enhancement after contrast injection	A heterogeneous solid cystic right frontal lesion in precentral gyrus with the cystic and hemorrhagic formation Thickening of the dura with avid enhancement A heterogeneous signal lesion with hemorrhage on T2-weighted images	Surgery	Improved symptoms without recurrence In six years
17562595	Musolino/ 2007 [12]	35/female	Cough, septic fever, decreased visual acuity in the left eye, and dysphagia	AIDS-related CNS lymphoma and pancytopenia (CD4+ lymphocyte count 10/µL, HIV-RNA viremia of 37000 copies/mL) Polyclonal hypergammaglobulinemia	Multiple bilateral cortical and subcortical solid lesions without either perifocal edema or cavitation	NM	Whole-brain radiotherapy with endovenous high-dose dexamethasone (20 mg daily)/ then two times weekly) Injections of intrathecal methotrexate (15 mg)	Death
17134026	Kaya/2006 [4]	18/m	Severe headache, dyspnea, fatigue, and anemia, Hepatomegaly	Idiopathic myelofibrosis	NM	T1-weighted images: homogenous meningeal enhancing after contrast	NM	NM

PMID	Author, Year	Age/Sex	Presenting Symptoms	Underlying Disease	CT Features	MRI Features	Treatment	Outcome
16440782	Shishime et al. (2005) [24]	63/m	NA	Idiopathic myelofibrosis	NA	NA	Whole-brain irradiation (30 Gy)	The lesion was controlled after one and a half years
15580341	Haidar et al. (200) [3]	9/f	Headaches unresponsive to routine treatment	Myelofibrosis	Bilateral high-signal soft tissue lesion with nodular appearance and homogeneous enhancement after contrast	T1- and T2-weighted images: dural hypoisignal lesions with enhancement after contrast Diffuse pachymeningeal thickening throughout both frontoparietal regions.	NM	NM
15461616	Ozdogu and Boga (2004) [25]	19/m	Headache, vomiting, and subconjunctival hemorrhage	Idiopathic myelofibrosis	Hyperdense and irregular meningeal lesion, which surrounded the brain with prominent homogenous contrast enhancement	Marked meningeal thickening	Radiotherapy	Partial regression of the lesion and symptomatic improvement
14707708	Ayyildiz et al. (2004) [26]	18/NM	Refractory headache, dyspnea, fatigue, and anemia	Idiopathic myelofibrosis	Intracranial meningeal diffuse mass	Diffuse signal enhancement surrounding the brain	Radiotherapy	Headache was slightly decreased MR after 5-month: symptoms had not resolved completely
14520481	Beckner et al. (2003) [27]	29/m	Known case of pilocytic astrocytoma in follow-up	Without any bone marrow disease or systemic EMH *EMH associated with pilocytic astrocytoma	NM	Enhancement of cystic tectal lesion is seen in T1-weighted image after tumor resection in three consecutive surgeries within five years with pilocytic astrocytoma microscopic features	Surgery	NM
8192010	Koch et al. (1994) [2]	69/m	Right-sided weakness, numbness, and hyperreflexia.	Myelofibrosis	NM	T1-w weighted image MR: a mass of intermediate signal intensity in the posterior part of the interhemispheric fissure T2-weighted image: Interhemispheric mass that is void of signal with perilesional edema in the adjacent parenchyma T1-weighted images MR after contrast: diffuse enhancement of mass and no enhancement of adjacent parenchyma. diffuse decrease In signal intensity throughout marrow of vertebral body, without abnormal enhancement or extradural masses	Preoperative transfusion of platelets Diagnostic craniotomy for biopsy of the largest lesion Whole-brain irradiation (2000 rad [20 Gy] in 10 fractions),	Marked symptomatic improvement Decrease in size of the lesions as shown by cranial MR imaging

PMID	Author, Year	Age/Sex	Presenting Symptoms	Underlying Disease	CT Features	MRI Features	Treatment	Outcome
8131005	Ohtsubo et al (1993) [28]	71/f	Hearing loss Left Hemiparesis Progressively spleen enlargement and pancytopenia	Postpolycythaemic myelofibrosis	Unenhanced CT: iso- or hyperdense broad base extra-axial masses Postcontrast scan: diffuse homogenous enhancement of the dural-based mass	NM	NM	Dead
1868657	Urman et al. (1991) [29]	39/m	i Ataxic gait for 10 days Disorientation in time and place Hyperactive DTR Bilateral extensor responses	Myelofibrosis	Irregular, lobulated, high-density, interhemispheric masses that extended along the falx cerebri and contained areas of lower attenuation centrally Tear-shaped masses on the coronal examination High-density thickening of tentorium bilaterally, which was relatively smooth in contour Diffuse low- attenuation of white matter bilaterally with linear sites of increased density in the corpus callosum and paramedian frontal lobes Following contrast infusion: the central area of lower density in the falx showed enhancement	NM	A single cranial dose of 600 rad	NM

PMID	Author, Year	Age/Sex	Presenting Symptoms	Underlying Disease	CT Features	MRI Features	Treatment	Outcome
3179950	Landolfi et al. (1988) [30]	62/f	High-grade fever and a progressive bilateral exophthalmos Neurologic signs: Right hemiparesis, episodic aphasia, and sphincter control loss	Polycythemia vera Secondary myelofibrosis	Multiple bilateral lesions with enhancement were seen around optic nerves Multiple extra-axial lesions were seen next to the inner table of the left hemicranium A marked increase in the thickness of the falx cerebri Both intraorbital and intracranial lesions	NM	Radiotherapy	Complete neurologic improvement after radiotherapy CT: subtotal improvement of the optic nerve and extra-axial lesions
3985737	Fucharoen et al. (1985) [31]	23/m	Generalized epileptic attacks, with prodromal bilateral hand jerky movement Loss of consciousness for five to ten minutes during the convulsion	Hemoglobin E-beta-thalassaemia	Hyperdense (60 Hounsfield) right frontoparietal inner table lesion with a peripheral irregular density	NM	Surgery Local irradiation treatment Phenobarbital sodium (60 mg/day)	Complete treatment after 3 month
6736389	Lund and Aldridge (1984) [32]	50/f	Frequent headaches for the past 2 weeks Sudden onset of expressive aphasia, lasting approximately 2h	Myelofibrosis	The mass over the left frontoparietal region measured approximately 5 cm at its base. It buckled the adjacent cerebral cortex with the obvious mass effect but produced minimal surrounding edema	NM	Follow-up and probable whole-brain radiation	NM

PMID	Author, Year	Age/Sex	Presenting Symptoms	Underlying Disease	CT Features	MRI Features	Treatment	Outcome
6633840	Gregorios et al. (1983) [33]	56/m	A 6-week history of memory disturbance, poor concentration, and personal change consisting of irritating, aggressive behavior, and tonic-clonic seizure	Malignant meningioma *EMH in a malignant meningioma	Large bifrontal tumor with the characteristics of falx meningioma and considerable both frontal lobes edema Frontoparietal edema midline shift from left to right and right-sided hydrocephalus	NM	Bifrontal craniotomy Medical postoperative course Cranial irradiation	Improved neurologically, but memory and effect remained imperfect
7093826	Kandel et al (1982) [34]	37/m	Difficulty in walking and spastic paresis of legs, urinary incontinence, headaches papilledema, and retinal hemorrhages,	Polycythemia vera	Midline vascular mass in falx cerebri with radiodensity approximates that of skull Large radiodense fusiform lesion involving the falx cerebri	NM	Dexamethasone plus cranial radiotherapy	Improvement in tumor size There was no neurological signs or symptoms in one year but after one year the patient died
7258183	Fucharoen et al. (1981) [13]	27/f	Dyspnea and chest pain, fever, and cough for four days	Hemoglobin E-beta-thalassaemia	NM	NM *Biopsy: solid, darkish red lesion was seen under dura mater, with attachment to the cerebral falx and dura Lesion extension was observed in the central part of the cerebral hemispheres with brain atrophy related to the lesion		Death seven days after admission

EMH: Extramedullary Hematopoiesis; M: Male; F: Female; NM: Not Mentioned; NA: Not Accessible; SDH: Subdural Hematoma; FLAIR: Fluid-Attenuated Inversion Recovery; SWI: Susceptibility-Weighted Imaging; DWI: Diffusion-Weighted Imaging. MRI: Magnetic Resonance Imaging, CT: Computed Tomography

Different treatment options used for these patients are hyper transfusion therapy with regular administration of hydroxyurea, low-dose radiotherapy, massive steroid treatment, and surgical resection of the hematopoietic mass (Table 1).

The emergence of refractory headaches with or without other signs of increased intracranial pressure in patients with relevant past medical history (mostly patients with myelofibrosis and thalassemia) should warrant consideration of the possibility of the presence of intracranial compensatory hematopoiesis. This could help earlier diagnosis and consequently, more successful treatment of these patien

Ethical Considerations

Compliance with ethical guidelines

All ethical principles are considered in this article. The participant was informed of the purpose of the research and its implementation stages. He was also assured about the confidentiality of his information, and if desired, the research results would be available to him. Written consent has been obtained from the subject. Principles of the Helsinki Convention were also observed. The Publisher has permission to publish the relevant Contribution.

Funding

This research did not receive any grant from funding agencies in the public, commercial, or non-profit sectors.

Conflict of interest

The authors declared no conflict of interest.

Acknowledgements

We would like to thank Erfan Mohammadi-Vajari for his contribution to this article and also the wonderful staff of the Radiology Department of Firoozgar Hospital.

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