Case 13036

Extramedullary hematopoiesis of the falx
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Section: Neuroradiology
Area of Interest: Neuroradiology brain Spine
Procedure: Diagnostic procedure
Imaging Technique: MR
Special Focus: Blood Case Type: Clinical Cases
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Patient: 63 years, female

Clinical History:

A 63 year old woman suffering from a mixed myeloproliferative syndrome for 14 years, with spleen and liver involvement, had a splenectomy and later signs of hepatomegaly and portal hypertension. She previously presented 3 episodes of transient ischemic attack, and recently presented with gait instability, and was therefore referred for brain MRI.

Imaging Findings:

Brain MRI was performed at 1.5T, mainly focused on the posterior fossa, searching for stroke or inner ear disease. Coronal T2WI showed a slightly high signal heterogeneous nodular lesion stretching on both sides of the falx, extending to the superior frontoparietal sulci. This lesion appeared as hyper signal on FLAIR, without mass effect. A strong and heterogeneous hyper signal T1 was seen after intravenous injection of gadolinium. The clivus and occipital condyles had a spontaneous low signal on T1 WI and a heterogeneous enhancement on fat saturated T1 WI. There was neither ventricular enlargement nor periventricular leucopathy.

A previous brain MRI had been performed to check a subdural hematoma seen on CT. The lesion was isosignal to white matter on T2 WI, a very low signal on SWI, a signal similar to the cortex on T1 WI, a heterogeneous low signal on DWI (b1000) and a heterogeneous high ADC value.

Discussion:

Extramedullary hematopoiesis (EMH) is described in patients suffering from bone marrow failure, especially in chronic anemia. This induces proliferation of hematopoietic cells outside the bone marrow, in the reticulo-endothelial system. The underlying cause is most often thalassemia in children and myelofibrosis in adults; the other causes are malignant hemopathies, hemoglobinopathies, infections and immune response [1, 2].

EMH mostly arises in the liver, the spleen, the lymph nodes, or in the mediastinum; it rarely affects the dura. When occurring in the dura, EMH is most of the time located in the spine, developing as masses leading to spinal cord compression, nerve root compression or mediastinal compression. Intracranial EMH is an even rarer entity, it may also arise in meningioma or in subdural hematoma. The clinical presentation of intracranial EMH is variable, from asymptomatic patients to intracranial hypertension or compression syndromes [3]. Meningeal EMH may present as headache, drowsiness, disorientation, obnubilation, epilepsy and hemiparesis. It has also been described as an incidental finding [4-9]. In our case the MRI features could not be related to the clinical symptoms of the patient, as there was no sign of cranial hypertension on MRI. Considering the past medical history of the patient, we reviewed the previous radiological examinations and found on a pancreatic MRI an enlargement of the vertebral gutter ribs, reinforcing the diagnosis of extramedullary haematopoiesis of the falx cerebri, associated with spinal extramedullary
Typical MRI appearance of meningeal EMH is extra-axial nodular tissue with well defined margins, that presents as isointense to gray matter on T1WI, hypointense to gray matter on fat sat T2WI, hypointense to gray matter on GRE, markedly hyperintense to gray matter on SWI, has no restricted diffusion, is hypointense to gray matter on unenhanced FLAIR, and presents strong enhancement on post Gd T1WI. Confirmation can be made either by biopsy or by Tc-99m sulfur colloid scan, which is a common marker of the reticuloendothelial system [6]. In this case further examinations were not asked by the oncologist.

The treatment is related to the systemic disease, moreover cerebral radiation therapy is very effective as hematopoietic (stem) cells are very sensitive to radiation. Surgery is less recommended as complete resection is difficult, due to the localisation and extension of intracranial EMH [10]. Both clinical data and radiological aspects are required to lead to that diagnosis.

**Differential Diagnosis List:** Regarding the whole file, we believe it is falx EMH, subdural hematoma, extramedullary hematopoiesis, meningioma, meningeal metastasis

**Final Diagnosis:** Regarding the whole file, we believe it is falx EMH.

**References:**


Description: Slightly high signal mass along the falx cerebri 

Origin: Department of Radiology, Institut Jules Bordet, Bruxelles, Belgium
Figure 2

**Description:** High signal lesion centered on the falx entering the sulci

**Origin:** Department of Radiology, Institut Jules Bordet, Bruxelles, Belgium
Figure 3

Description: Diffuse and intense high signal of the lesion  

Origin: Department of Radiology, Institut Jules Bordet, Bruxelles, Belgium
Figure 4

Description: Osseous hyposignal within the clivus and occipital condyles

Origin: Department of Radiology, Institut Jules Bordet, Bruxelles, Belgium
Description: Heterogeneous enhancement of the clivus and occipital condyles

Origin: Department of Radiology, Institut Jules Bordet, Bruxelles, Belgium
Figure 6

Description: Strong low signal lesion

Origin: Department of Radiology, Institut Jules Bordet, Bruxelles, Belgium
Description: Vertebral gutt enlargement  
Origin: Department of Radiology, Institut Jules Bordet, Bruxelles, Belgium