CORRESPONDENCE

Seizures as the initial symptom of a diffuse dura mater and choroids plexus haematopoiesis revealing myelofibrosis

A 63-year-old woman was admitted with a one-month history of visual loss, headaches and seizures. On initial examination, she was confused with bilateral papilloedema, enlarged liver and spleen, associated with cutaneous purpura. Laboratory examinations revealed pancytopenia: haemoglobin level was 9.5 g/dL, white blood cell count 4500 per millimetre cube and platelet count 65,000 per millimetre cube. A peripheral blood smear revealed anisocytosis, poikilocytosis, teardrop cells, and leukoerythroblastic changes. Brain MRI revealed an irregular, lobulated thickening of the dura mater, surrounding the brain and extending along the falx cerebri (Fig. 1A–D). The meningeal thickening was homogeneous, isointense on T1-weighted images (WI), hyperintense on T2-WI with a striking enhancement after Gd-DTPA injection. Similar changes were observed in the enlarged choroid plexus. They were surrounded by an intraparenchymatous signal change, involving the deep subcortical white matter and sparing the grey matter. Bone marrow aspiration and biopsy revealed fibrosis, focal hypercellular marrow, megakaryocytic hyperplasia and myeloid metaplasia, thus permitting the diagnosis of myelofibrosis. A meningeal biopsy and an indium-111 chloride marrow scan showed intense uptake of activity in the dura mater and choroid plexus, confirming the diagnosis of extramedullary haematopoiesis (EMH). Neurological symptoms regressed after high-dose corticotherapy associated with hydroxyurea.

EMH is a common consequence of myelofibrosis. In adult mammals, haematopoiesis normally occurs in the

Figure 1   MRI: axial T2- (A) and T1-WI (B); axial (C) and coronal (D) T1-WI after injection of contrast media. Homogeneous, diffuse irregular and lobulated thickening of the dura mater, extending along the falx cerebri, isointense on T1- and hyperintense on T2-WI. There was a marked enhancement after injection. Choroid plexus appeared bilaterally enlarged, with the same pattern of signal and enhancement, surrounded by a parenchymal hypersignal, involving the periventricular white matter. This oedema, which could explain headaches and seizures, have been due to either a mass effect or pathologic venous drainage. IRM, coupes axiales en séquences pondérée T2 (A), T1 (B), axiale (C) et coronale (D) T1 après injection de chélate de gadolinium. Épaississement diffus, homogène, irrégulier, plurilobé de la dure-mère, en isosignal T1, hyperintense en T2, se rehausssant de manière intense. Les plexus choroides sont hypertrophiques, hypervasculaires. On note une anomalie de signal en T2 du parenchyme cérébral adjacent, affectant la substance blanche périventriculaire. Cet œdème, qui pourrait expliquer céphalées et crise, peut être la conséquence de l’effet de masse de la pachyméningite ou d’une anomalie du drainage veineux des hémisphères.
Correspondence

bone marrow, which supports simultaneously the life-long maintenance of stem cells and the regulated production of end-stage lymphoid, myeloid and erythroid cells. When blood availability is affected, haematopoiesis can be reactivated as a compensatory mechanism in organs where it previously occurred during embryonic life [1]. Intracranial EMH has rarely been reported and most frequently invades the cranial dural, the optic nerve sheath and the diploic space of the skull [1,2]. Although asymptomatic EMH has been reported [2], headaches and seizures are the main clinical symptoms. Dural EMH has been described as multiple contrast-enhanced extra-axial masses. Moreover, the present case suggests that the choroid plexus also have the capacity to reactivate haematopoiesis expression after a period of latency under pathologic circumstances, such as myelofibrosis. The ability of choroid plexus to generate blood, described in animal models [3,4], could be explained either by the reactivation of a gene programme in differentiated cells that have retained the capacity to reacquire an embryonic phenotype, or more likely, by the differentiation of resident precursor cells with haemovascularogenic potential when the physiological conditions require it to maintain homeostasis. In conclusion, we here reported a rare case of a choroid plexus location of EMH.

References


O. Naggara * 
E. Meary
R. Marsico
C. Oppenheim
J.-F. Meder
Department of Neuroradiology, centre hospitalier Sainte-Anne, Descartes University, Paris-5, 1, rue Cabanis, 75013 Paris, France
*Corresponding author.
E-mail address: o.naggara@ch-sainte-anne.fr (O. Naggara).
Available online 12 August 2008

© 2019 Elsevier Masson SAS. Tous droits réservés. - Document télé chargé le 13/05/2019 Il est interdit et illégal de diffuser ce document.