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Neurovascular disease

Subarachnoid and intra-cerebral hemorrhage in young adults: Rare and underdiagnosed



Hémorragie cérébro-méningée de l'adulte jeune : diagnostics rares et sous-estimés

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ABSTRACT

Objectives. – Convexity subarachnoid and intra-cerebral hemorrhages, in patients aged < 50 years, are always a diagnostic challenge. This condition is characterized by acute headaches with or without neurological symptoms and/or seizures, and by the radiological demonstration of subarachnoid and/or intra-cerebral hemorrhages and, more rarely, by the association of ischemic events.

Patients and methods. – In a prospective series of 30 consecutive patients (median age 31 years; 22 women) with a subarachnoid and intra-cerebral hemorrhages, 19 were diagnosed with reversible cerebral vasoconstriction syndrome (RCVS), 7 with cerebral venous sinus thrombosis (CVST), and 4 with a bleeding mycotic aneurysm (MA).

Results. – RCVS appeared spontaneously in 16 patients and was related to the postpartum period in three cases. Subarachnoid hemorrhage (SAH) was demonstrated in 24 patients as follows: 18 cases were in cortical areas, 4 were in the polygon of Willis, one was inter-hemispheric, and one was inter-hemispheric/intra-cerebral. A convexity pure intra-cerebral hemorrhage (ICH) was recorded in 6 cases. Among the 7 patients suffering from CVST, the superior sagittal sinus was involved in 4 cases, the transverse sinuses (TS) in 2, and the TS plus sigmoid sinus (SS) in one.

Conclusion. – The three most common causes in this series were RCVS, followed by CVST and bleeding from MA. Because of atypical clinical or radiological presentations, this large spectrum of etiologies can cause diagnostic difficulties. Therefore, careful analysis is needed to ensure correct and prompt diagnosis and to avoid any dangerous delays in management.

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R É S U M É

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Thrombose veineuse cérébrale

Introduction. – Les hémorragies cérébro-méningées de la convexité chez les patients de moins de 50 ans constituent souvent un défi sur le plan étiologique. Ces entités sont caractérisées par l'association de céphalées sévères avec ou sans symptômes neurologiques et/ou épilepsie, et par la preuve radiologique d'une hémorragie méningée et/ou intracérébrale et, plus rarement, par l'association d'évènements ischémiques, lesquels sont souvent trompeurs.

Patients et méthodes. – Les auteurs décrivent une série prospective de 30 patients consécutifs, hospitalisés pour hémorragie cérébro-méningée en urgence en neurochirurgie pendant 18 mois. Dix-neuf ont été diagnostiqués comme angiopathie aiguë réversible (AAR), 7 comme thrombose d'un sinus veineux cérébral (TSVC) et 4 comme saignement d'un anévrisme mycotique (AM).

Résultats. – Il y avait 22 femmes et 8 hommes avec un âge médian de 31 ans. L'AAR est apparue sans contexte particulier chez 16 patients, et par contre est survenue en période postpartum dans 3 cas. Aucune condition spécifique n'a été observée chez les patients souffrant de TSVC et d'AM. Une hémorragie méningée a été démontrée chez 24 patients avec pour localisation : la convexité (18 cas), la base du crâne (4 cas), la région interhémisphérique (1 cas), la région interhémisphérique et intracérébrale (1 cas). Une hémorragie intracérébrale pure de la convexité a été retrouvée dans les 6 cas restants. Parmi les 7 patients souffrant de TSVC, le sinus sagittal supérieur était incriminé dans 4 cas, les sinus transverses (ST) dans 2 cas, le ST et le sinus sigmoïde dans un cas.

Conclusion. – Les trois causes les plus fréquentes d'hémorragies cérébro-méningées de la convexité dans cette série étaient l'AAR, suivie par la TSVC et par le saignement d'un AM. Devant des tableaux clinico-radiologiques atypiques, ce large spectre d'étiologies peut constituer une difficulté diagnostique. Elles doivent être soigneusement recherchées pour permettre un diagnostic rapide et éviter un dangereux retard de prise en charge.

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1. Introduction

Subarachnoid and intra-cerebral hemorrhages in young patients are always a diagnostic challenge: prompt identification of its peculiar underlying pathologies is crucial to avoid potentially harmful examinations and management therapies. Subarachnoid hemorrhage (SAH) can be a possible confusing and misleading feature of reversible cerebral vasoconstriction syndrome (RCVS) as well as of mycotic and/or berry aneurysms. In the present study, the authors report their experience on a series of 30 patients managed for RCVS, cerebral venous sinus thrombosis (CVST), and mycotic aneurysm (MA), highlighting the clinical and radiological features that should be considered to enable prompt and correct diagnosis and management.

2. Patients and methods

This prospective single-center study contained 30 consecutive young adults (< 50 years old) observed for SAH and convexity intra-cerebral hemorrhage (ICH) in Paris during an 18-month period (January 2007–June 2008). The authors had chosen to study patients under 50 years because the frequency of ICH related to high blood pressure and cerebral amyloid angiopathy is less [1,2]. During this period, our center managed 159 emergency subarachnoid and intra-cerebral

hemorrhages of which 101 were polygon of Willis aneurismal hemorrhages, 18 were arteriovenous malformations hemorrhages, and 30 were related to other pathologies related to the topic of this report (18.9% of all subarachnoid and intra-cerebral hemorrhages). All data (gender, age, biological analyses, headache characteristics, past medical histories, recent pregnancy and delivery, medications, and drugs taken within 15 days) were analyzed and prospectively used to build a database.

Headache was evaluated by a verbal scale, ranging from 0 (no pain) to 10 (maximum pain). Biological analyses were made for all patients (100%), including full blood counts, electrolyte levels, liver- and renal-function tests, C-reactive protein (CRP) levels, erythrocyte-sedimentation rate (ESR), rheumatoid factors, antinuclear and anti-neutrophil-cytoplasmic antibody tests, blood and urine tests for cannabinoids, cocaine and amphetamine, a test for vanillylmandelic acid, and blood cultures. In addition, four selected patients received a cardiac ultrasound to explore/confirm endocarditis. All patients underwent an emergency brain and angio-CT-scan, as well as brain magnetic resonance imaging (MRI) with fluid-attenuated inversion recovery (FLAIR) images. Six (20%) patients had an additional lumbar puncture and all 30 patients had a cerebral digital subtraction angiography (DSA). All patients also underwent clinical and radiological DSAs during the following 12 weeks. A MRI/magnetic resonance angiogram (MRA) scan was conducted every 6 months for up to 1 year, as applicable.

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