Giant Arachnoid Granulation (GAG) in a child with acute headache

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Introduction
The cerebrospinal fluid is normally reabsorbed by arachnoid villi or granulations of Pacchioni that allow the flow in the venous sinuses, especially in the superior sagittal sinus. Only the largest Pacchioni granulations, however, come directly into the lumen of the sinuses with the smaller extend across the subdural space (1). We refer to arachnoid granulations as “giant” (GAG) when they have such dimensions as to fill the lumen of the venous sinus and cause local dilation and filling defects (Kan et al. 2). Rarely, this condition causes clinical symptoms, especially a clinical picture suggestive of intracranial hypertension, however, this finding implies serious problems of differential diagnosis with other pathological conditions (cerebral venous thrombosis, neoplasms, etc.) (1). We present a case of a 10 years old patient with progressive headache and a GAG involving his transverse venous sinus.

Case report
A 10 year-old boy came to our observation for the second episode of acute headache associated with fever and left deviation of buccal rhyme. Physical examination appeared to be normal. The neurological and neuropsychological assessments excluded the presence of focal signs. Routinely blood tests and coagulation profile study resulted to be normal. Neuro-radiological investigations (CT, MRI and Anglo-CT) showed, in the context of the gulf of right jugular, an ovoid 14.8 x 7.4 mm mass, not clearly interpretable (arachnoid granulation?, intradural thrombus?, glioma tumor?). Suspecting a cerebral venous thrombosis, antithrombotic treatment (enoxaparin sodium) was started, with regression of clinical symptoms. A central nervous system-MRI scan was performed again after few days of treatment. It demonstrated the filling defect without showing significant changes in the size of the lesion, thus the diagnosis of giant arachnoid granulation was confirmed. The mass was peripherally iso-intense and centrally hypo-intense to brain parenchyma with focal central calcifications. The neurological and neuropsychological assessments excluded the presence of focal signs. The child, however, started folic acid therapy.

Discussion
Arachnoid granulations or villi are growths of arachnoid membrane into the dural sinuses, through which cerebrospinal fluid (CSF) enters the venous system from the subarachnoid space. The growth of arachnoid membrane into the dural sinus was first described by Pacchioni in 1705 (3). These projections are called arachnoid villi or arachnoid granulation (AG), depending on their size. Arachnoid villi are microscopic, whereas granulation is visible to the naked eye. AGs are absent at birth and develop in infants at the time of closure of the fontanels and increase with age, in numbers and size, in response to increased CSF pressure from the subarachnoid space and are usually quite evident by 4 years of age (4). AGs, in general are a rare polymorphism, with normal levels of serum homocysteine. The child, however, involved in homocysteine metabolism. Homocysteines have more than 50% reduced enzyme activity, but the effect of impaired MTHFR function on homocysteine levels is dependent on folate intake (10). The classic MTHFR C677T gene polymorphism is weakly associated with an increased risk of VTE (Venous thromboembolism).

The relative risk for venous thromboembolism, due to MTHFR decreased activity, is slightly increased when a condition of double heterozygosis is present. However, our patient started folic acid therapy.

We experienced a case of a giant arachnoid granulation misdiagnosed as dural sinus thrombosis. Before diagnosing the sinus thrombosis, giant arachnoid granulation should be considered in differential diagnosis. MRI is the most useful tool to differentiate giant arachnoid granulation from dural sinus thrombosis.

Fig 1: Anglo-CT finding

References
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variant presenting as a mass within the dural venous sinus Am J neuroradiol 1995; 16:901-04