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Case Report

An unusual vermiform giant arachnoid granulation

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ABSTRACT

Arachnoid granulations are outpouchings of arachnoid membrane which extend into the dural sinuses or calvarium, surrounded by a capsule of dense connective tissue. Within dural sinuses, these appear as well-defined, nodular, rounded, or ovoid structures of focal localization. However, it is important to be aware of their variability in presentation in order to correctly identify them and distinguish them from other dural sinus pathology, especially a misdiagnosis of venous sinus thrombosis with risks of unnecessary anticoagulation, intravascular thrombolysis/thrombectomy, or invasive intracranial pressure monitoring. Here we demonstrate a case of a previously unreported giant intrasinus arachnoid granulation of an unusual vermiform morphology, unduly elongated up to 6 cm in length, involving a significant segment of the superior sagittal sinus. The proof of this diagnosis was the radiologic appearance on multiple modalities and an unchanged appearance over the long-term.

Introduction

Arachnoid granulations are subarachnoid space outpouchings into the dural sinuses or calvarium that allow for the absorption of the cerebrospinal fluid into the venous system [1,2]. While inconspicuous at birth, arachnoid granulations seem to present over time, becoming obvious by 18 months to 4 years of age [1]. Their characteristic distribution within the sinuses predominantly involves the superior sagittal, transverse, and straight sinuses [3]. Moreover, it has been described that veins are usually present at the periphery of small arachnoid granulations and in the center of large ones [4]. Arachnoid granulations can vary in size and enlarge with increase in pressure in the subarachnoid space [1,2,5]. They are termed “giant” when they are large enough to occupy or cause expansion of the lu-

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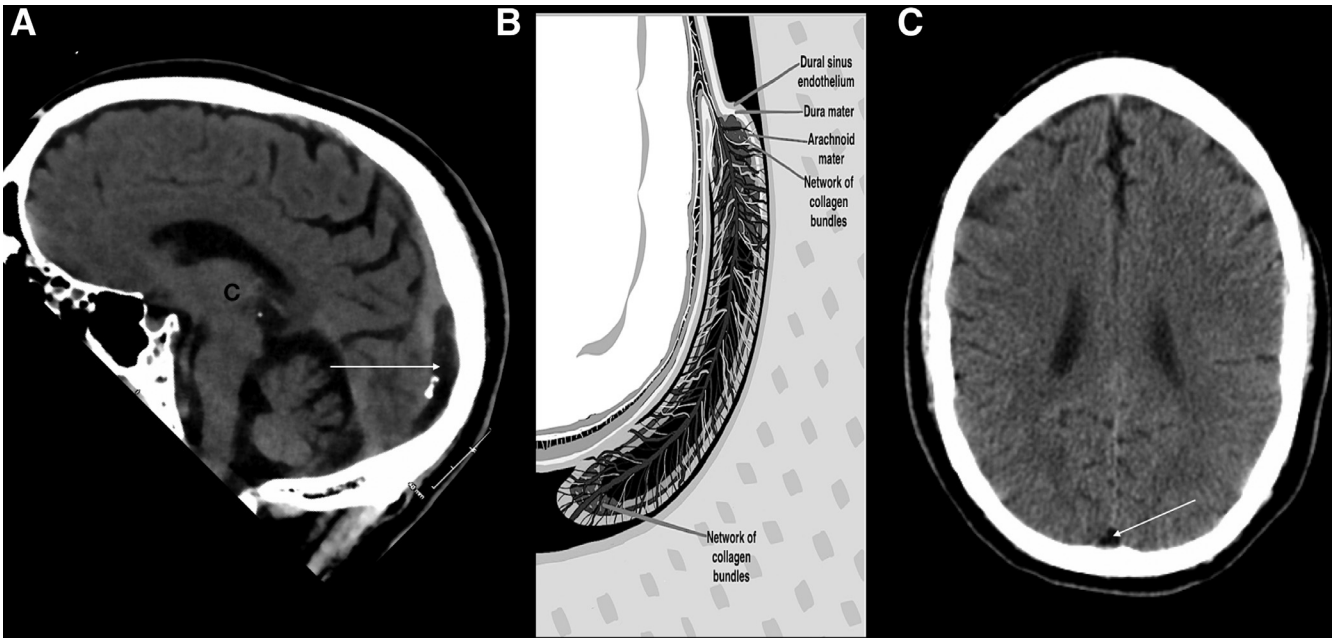

Fig. 1 – (A) Sagittal midline CT slice shows a hypodense (˜CSF density) arachnoid granulation in the posterior aspect of the superior sagittal sinus (arrow). (B) Schematic diagram depicting the typical internal structure of an arachnoid granulation, with the elongated morphology and superior pedicle described in this case incorporated (illustration by author TM). (C) Noncontract CT from 8 years previously demonstrating identical lesion within the superior sagittal sinus (arrow).

men of a dural sinus [5], or sometimes defined as greater than 1 cm in diameter [6].

Most intrasinus giant arachnoid granulations are focal, well-defined nodular structures that take a round or oval shape and range from a few millimeters to a few centimeters in size [4,7]. Here we describe an unusual case of a giant arachnoid granulation within the superior sagittal sinus that involves a significant segment of the dural sinus, is of a larger size and more elongated than intrasinus giant arachnoid granulations previously described in literature.

Case report

A male patient in his 50s presented to our emergency department with acute symptoms of altered mental status of unclear etiology. A noncontrast computed tomography (CT) of the head was performed to rule out intracranial hemorrhage and revealed a tubular elongated lesion within the posterior superior sagittal sinus (Fig 1A). Hounsfield units within this lesion matched cerebrospinal fluid (CSF) (Fig 2 F). An magnetic resonance (MR) was also performed and the lesion matched the appearance of CSF, that is hyperintense on T2-weighted MR images (Fig 2 C), and hypointense without appreciable enhancement on T1-weighted imaging (Fig 2 E). The expected T2-hypointense flow void above the lesion and the entry of the vascular bundle into the neck of the superior end of the lesion was also demonstrated on MR (Fig 2 A,B). T2-weighted FLAIR imaging showed that the lesion exhibited suppression of signal similar to CSF (Fig 2 D). Furthermore, a noncontrast CT head of this patient taken 8 years prior confirmed the presence of a lesion of similar shape, size, and attenuation in the posterior superior sagittal sinus (Fig 1 C).

The long-standing giant arachnoid granulation was unrelated to the patient's acute change in mental status. Following neurology consultation, the patient's medication dose of carbidopa/levodopa for Parkinsonism was adjusted to a lower daily intake. The patient showed marked improvement in mental status in one week and was discharged.

Discussion

Arachnoid granulations are arachnoid membrane outgrowths extending into the dural sinuses or calvarium that allow for the absorption of the cerebrospinal fluid from the subarachnoid space into the venous system [1,2]. They were first characterized by Antonio Pachionni in 1705 and given their original name of "Pachionni granulations" [8]. Larger intrasinus arachnoid granulations are commonly termed "giant" arachnoid granulations and their shape has been well described in the recent literature. They have been identified as forming mostly focal, well-defined, nodular entities of round, or oval shape [4,7].

It has been previously thought that arachnoid granulations could be distinguished from dural sinus thrombosis based on the fact that arachnoid granulations produced focal, well-defined, nodular defects within the sinuses, whereas thrombosis extended to involve a larger segment of a sinus [9,10]. However, our finding of a 6 cm tubular giant arachnoid granulation that occupied a large segment of the superior sagittal sinus has demonstrated that this distinction might not be as

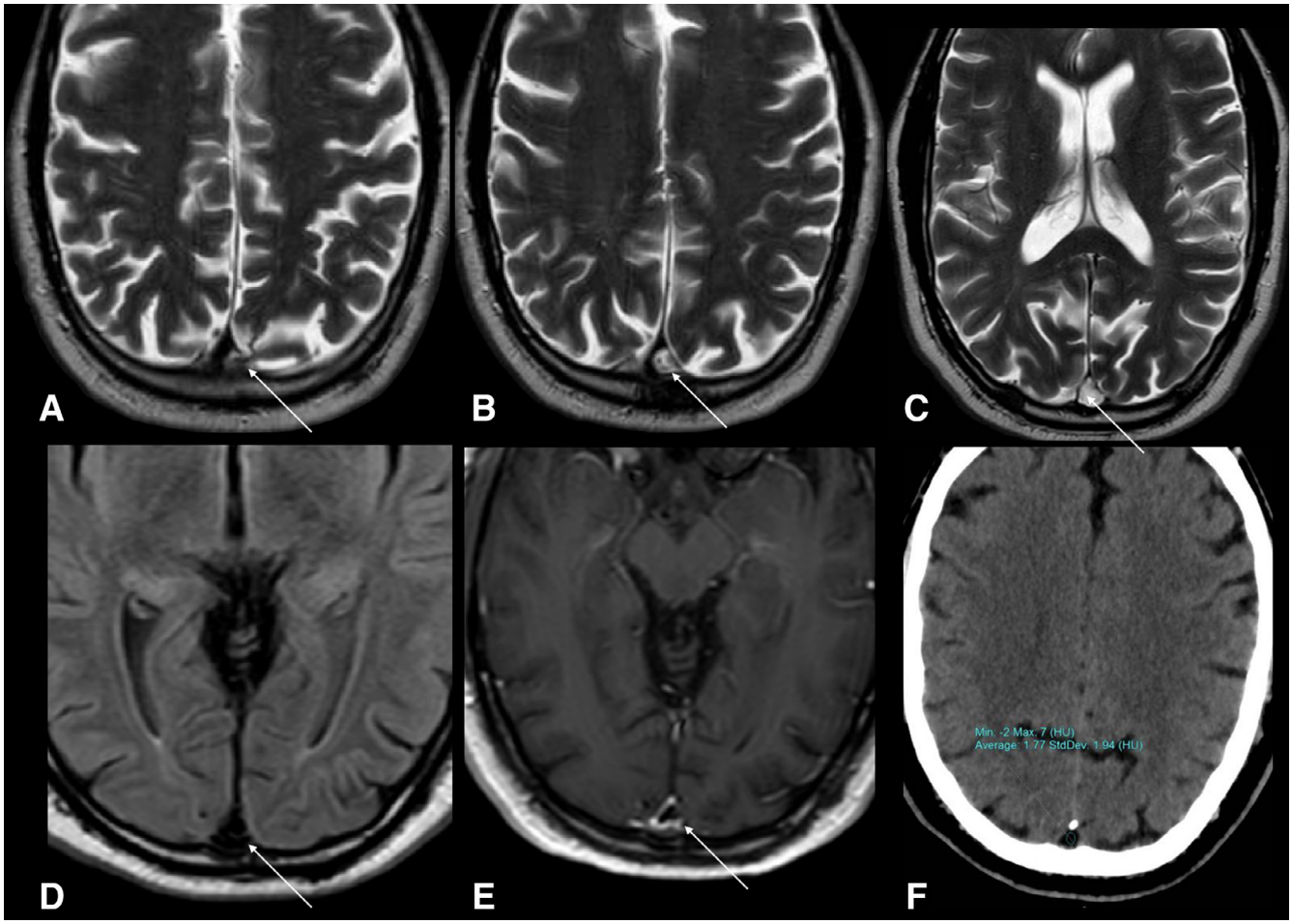

Fig. 2 – Top row: T2-weighted axial slices, ordered craniocaudally, demonstrating normal flow void in the superior sagittal sinus above the lesion (A), vascular pedicle of the arachnoid granulation entering the sinus (A and B), and CSF-isointense arachnoid granulation (C) obliterating any appreciable flow void within the superior sagittal sinus. Bottom row: T2-weighted FLAIR (D) shows suppression of signal similar to CSF, and postcontrast T1-weighted imaging (E) shows a nonenhancing T1-hypointense filling defect. Axial noncontrast CT (F) demonstrates CT density similar to CSF.

straightforward only based on shape differences. Beyond the mere presence of the filling defect, however elongated, careful attention should be paid to comparing the intensity of the giant arachnoid granulation to the intensity of the CSF on T1-weighted imaging, T2-weighted imaging, and CT density. This includes noting its suppression on T2-weighted-FLAIR imaging, observing the degree of its enhancement on a postcontrast study, and quantifying its Hounsfield units (HU) on a noncontrast CT as necessary adjuncts to distinguish it from dural sinus thrombosis. Dural sinus thrombosis often exhibits hyperintensity on T1-weighted imaging and hyperdensity on CT in early evolution [10], whereas arachnoid granulations typically present as lesions that are hypodense on CT (isodense to CSF), and often isointense to CSF on multiple MR sequences and lack of solid enhancement or hypointense blooming susceptibility artifact. Some exceptions to these characteristics are attributable to CSF flow artifacts or connective tissue/vascular content within the arachnoid granulation [6].

Some variation in shapes of the giant arachnoid granulations could potentially be explained by their architecture. It has been demonstrated that lining the pedicle, body, and apex, which are the components of an arachnoid granulation, is a layer of connective tissue comprised of bundles of collagen fibers that originate from the dura mater and the granulation itself [4,9,11]. Furthermore, the composition of this layer can vary across the arachnoid granulation [11]. In addition, several studies revealed that arachnoid granulations contain elastic collagen fibers within their core that play a role in determining their shape in accordance with the developmental stage of the granulation [4,11,12]. The proportion of elastic collagen fibers contained within a granulation and the compliance of the connective tissue layer that lines it on the outside could potentially determine the shape, the arachnoid granulation eventually takes [4]. We believe that the giant arachnoid granulation that we have observed might belong to the similar group of few arachnoid granulations that exhibit an unusual shape and could potentially exemplify a unique architecture that is different from the more typical and easily recognized round or ovoid ones, especially the unusual large size and elongation of the giant arachnoid granulation that in our patient. Other po-

tential etiologies we suggest for the unique morphology of the granulation in the current study include adhesions between the apex of the granulation and the dural sinus wall early in development, with resultant lengthening as the sinus grew, or further elongation of an ovoid granulation in the downstream direction through long-standing hemodynamic effects of blood flow around the granulation over this long segment and converging below it.

Although the association of arachnoid granulations with patient symptoms remains unclear [13,14], the importance of distinguishing arachnoid granulations from dural sinus pathology is essential in making the right diagnoses and preventing inappropriate therapy. Arachnoid granulations are considered incidental findings that represent a normal anatomic entity commonly found in the adult population [15]. However, they can closely mimic pathology within the dural sinus, most importantly and commonly dural sinus thrombosis, as well as less common lesions such as sinus cavernoma, tumors such as meningioma or metastasis invading the sinus [2,7,10,13]. If an arachnoid granulation is mistakenly diagnosed for dural sinus thrombosis, the patient could be started on anticoagulation therapy, with potentially serious and unnecessary consequences, including intracranial bleeding [16–18], which can result in devastating sequelae for the patient, including requiring additional medical therapy, affecting a patient's ability to work, impacting major organ systems, and even resulting in fatality [18]. In a study by Linkins et al the authors examined 4374 patient-years of oral anticoagulant therapy from 33 studies and determined that major bleeding had a 13.4% case-fatality rate (95% confidence interval, 9.4%-17.4%) and intracranial bleeding had 1.15 per 100 patient-years rate (confidence interval, 1.14-1.16 per 100 patient-years) [17]. Moreover, the nonbleeding risks of anticoagulation therapy, such as vascular calcification, osteoporosis, anticoagulation-related nephropathy, and high costs of the drugs [18] are factors that could produce a considerable impact on a patient's quality of life.

Conclusion

It is important to be aware of unusual morphologies of intrasinus arachnoid granulations. An unusually elongated filling defect within a dural sinus on venograms, or venogram like postcontrast-imaging studies, could still represent an atypical morphology of a normal arachnoid granulation. This distinction can be established by meticulously considering all other CT and MR imaging characteristics of arachnoid granulations in terms of internal density and signal, and avoid expensive and potentially harmful effects of additional treatment or workup.

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