

Total Agenesis of Superior Sagittal Sinus and Falx Cerebri in a Patient Who Has a Subacute Subdural Hematoma Crossing Midline: Case Report

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Received, August 26, 2012.

Accepted, December 11, 2012.

Published Online, January 10, 2013.

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BACKGROUND AND IMPORTANCE: Anatomic variations of the superior sagittal sinus (SSS) and falx cerebri (FC) are uncommon in that agenesis of these structures is extremely rare. We report an extremely rare anatomic variation, total agenesis of the SSS and FC, and briefly discuss it from the anatomical, embryological, radiological, and clinical perspectives.

CLINICAL PRESENTATION: A 49-year-old woman presented with long-standing headache, gait disturbance, and nausea. Imaging studies showed a bilateral subdural hematoma crossing the midline, dilated venous structures, and perineural cysts, but SSS and FC. Following right-sided hemiparesis and consciousness disturbances, the subdural hematoma was evacuated from a left-sided parietal burr hole because of thick hematoma in this side. After the surgical evacuation, the hemiparesis and consciousness disturbances were regressed; however, she still had severe headache. On account of ongoing headaches and related imaging findings, it was thought that she had possible spontaneous intracranial hypotension. She was treated with autologous epidural blood patch and recovered well.

CONCLUSION: Agenesis of the SSS and FC are extremely rare variations. Agenesis of the SSS results in development of alternative venous pathways and may lead to misdiagnosis as dural arteriovenous fistulas. Agenesis of FC may cause diagnostic confusion, because subdural pathologies such as hematomas can cross the midline in rare occasions.

KEY WORDS: Absence, Agenesis, Falx cerebri, Spontaneous intracranial hypotension, Subdural hematoma, Superior sagittal sinus

Neurosurgery 72:E863–E867, 2013

DOI: 10.1227/NEU.0b013e318285ba68

www.neurosurgery-online.com

Anatomic variations of intracranial dural venous sinuses are commonly encountered in practice and most frequently involve the transverse sinuses.¹ Anatomic abnormalities of the superior sagittal sinus (SSS) are uncommon and mostly include duplication of the rostral SSS, unilateral hypoplastic rostral SSS, complete hypoplastic rostral SSS, hypoplastic dorsal SSS, and atresia of the rostral SSS.^{2–5} However, agenesis of the SSS is such a rare abnormality that we were unable to find any reference in the English literature.

Abnormalities of the falx cerebri (FC) are also infrequent like SSS and mostly seen as fenestra-

tion, hypoplasia, or agenesis with involvement of only the anterior part of the FC.^{6,7} Complete agenesis of the FC is very rare as is that of the SSS and has not been reported previously.

In this article, we present an extremely rare anatomic variation, total agenesis of the SSS and FC, in a patient who possibly has intracranial hypotension with an unusual subdural hematoma that crosses the midline. Anatomy and embryologic development of the SSS and FC, imaging findings, and related consequences of the total agenesis of these structures are briefly discussed.

CASE HISTORY

A 49-year-old woman who had been reporting headaches for 2 weeks was referred to the emergency department owing to an exacerbation

ABBREVIATIONS: DSA, digital subtraction angiography; FC, falx cerebri; SSS, superior sagittal sinus

of the long-standing headaches and additional complaints including gait disturbance and nausea. The neurological or developmental history of the patient was unremarkable. There was a lack of familial history related to her condition. A computed tomography (CT) scan of the brain was performed initially in the emergency setting and showed bilateral subdural hematoma with multiple membranous septa (Figure 1). A magnetic resonance (MR) examination was done subsequently and revealed rather unusual subdural hematomas joining each other at the midline owing to absence of the FC by which bilateral cerebral hemispheres were completely surrounded (Figure 1). In addition, an MR scan indicated tight posterior fossa with cerebellar tonsillar herniation, compression of corpus callosum by subdural hematoma, interdigitation of the cerebral gyri at the midline, a rather large deep

vein (tentorial sinus), prominent thickening of the supra- and infratentorial dura, and normal formation of the tectum and tentorium (Figure 1). On account of the existence of the dilated tentorial sinus, a digital subtraction angiography (DSA) was performed to exclude a potential arteriovenous malformation; it showed complete absence of all parts of the SSS, inward displacement of the venous structures owing to the subdural hematoma, and prominent tentorial sinus, but no arteriovenous fistula (Figure 2). There was no distinct unilateral source for hemorrhage. After the DSA, a MR myelogram and whole-spine MRI using short-inversion–time-inversion recovery sequences were done because suspected intracranial hypotension showed multiple perineural cysts at virtually all spinal cord levels, especially at the sacral levels (Figure 3). MR cisternography with

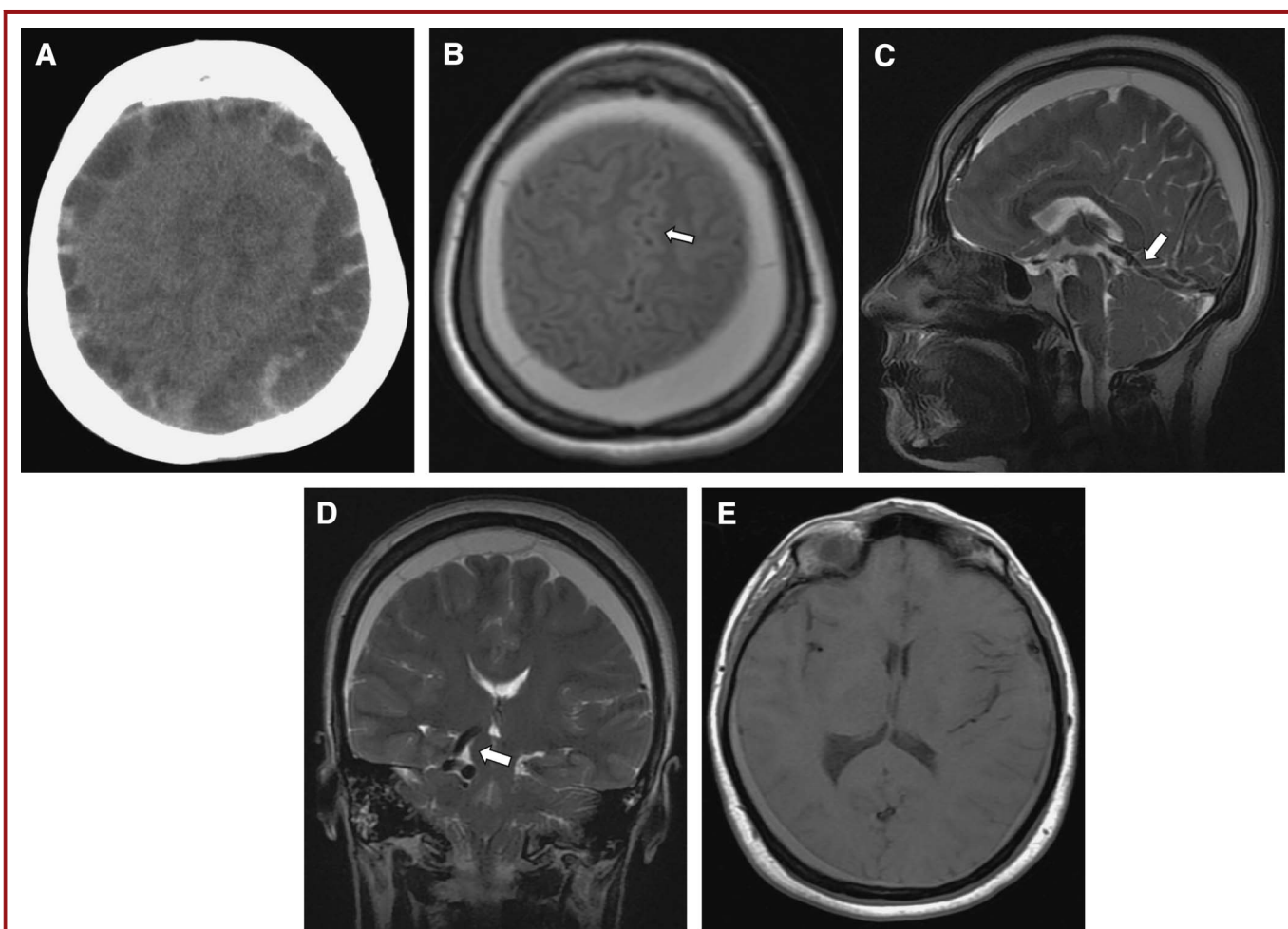


FIGURE 1. **A**, axial nonenhanced CT image shows bilateral subdural hematoma with multiple membranous septa. **B**, axial T2W FLAIR image depicts subdural hematoma surrounding the brain parenchyma and joining at the midline because of lack of FC. Note the interdigitation of midline gyri (arrow). **C**, sagittal T2W image demonstrates lack of the SSS, subdural hematoma, prominent tentorial sinus (arrow), compression of the cerebellar hemisphere by subdural hematoma, tight posterior fossa with cerebellar tonsillar herniation, and normal configuration of tectum and tentorium. **D**, coronal T2W image shows bilateral subdural hematoma surrounding the brain parenchyma joining each other at the midline, compression of the corpus callosum and cerebellar hemispheres. Also note the prominent right tentorial sinus (arrow). **E**, axial T1W image demonstrates the presence of 2 separate lateral ventricles. FLAIR, fluid-attenuated inversion recovery; FC, falx cerebri; SSS, superior sagittal sinus.

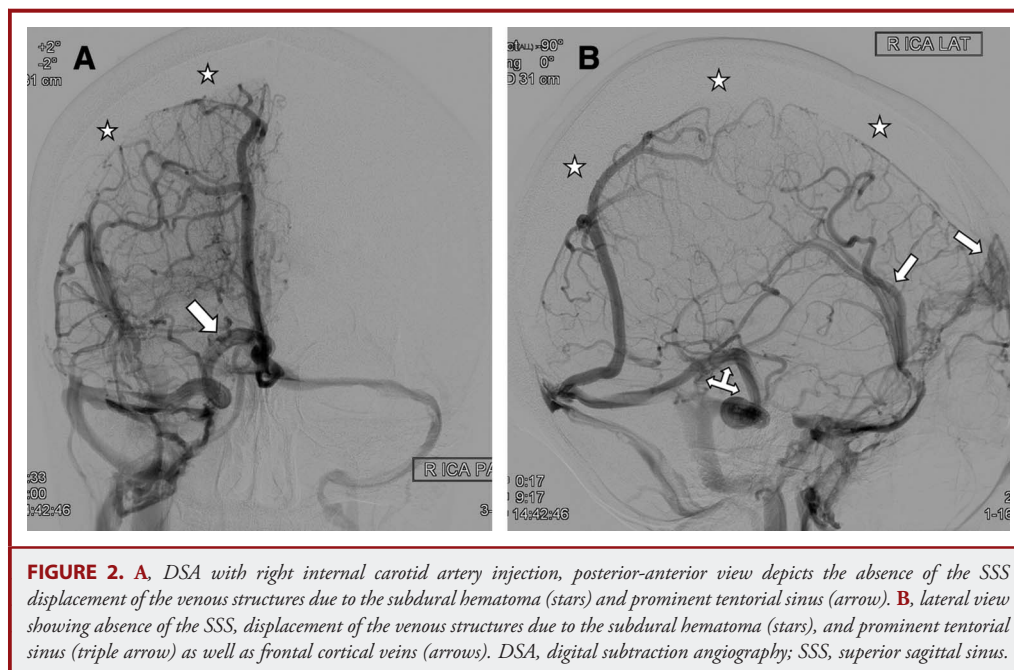


FIGURE 2. A, DSA with right internal carotid artery injection, posterior-anterior view depicts the absence of the SSS displacement of the venous structures due to the subdural hematoma (stars) and prominent tentorial sinus (arrow). B, lateral view showing absence of the SSS, displacement of the venous structures due to the subdural hematoma (stars), and prominent tentorial sinus (triple arrow) as well as frontal cortical veins (arrows). DSA, digital subtraction angiography; SSS, superior sagittal sinus.

contrast media was not performed because of cerebellar tonsillar herniation and tight posterior fossa to prevent further probable herniation. On the first day, the patient's clinical condition and vital signs were stable while she was under medical treatment and observation in the emergency department. On the second day, the patient suddenly complained of more intense headaches, vomiting, diplopia, photophobia, and right-sided slight hemiparesis, followed by decreased consciousness. Repeated CT scan showed worsening of the initial findings. She was taken to the operating room for surgical evacuation of subdural hematoma through a left-sided parietal burr hole because of the thick hematoma in this side. After surgery, hemiparesis and consciousness disturbances were regressed, but she still had a severe headache. Because of the ongoing bilateral subdural hematoma on control imaging studies, we accepted that it was probably secondary to intracranial hypotension syndrome. Therefore, the patient was treated with an autologous epidural blood patch at the L2-L3 intervertebral level 1 day after surgery; she improved back to her normal neurological baseline within 2 days. She was still asymptomatic and able to walk without gait disturbance at her 2-month clinical follow-up. Control CT performed 4 weeks after epidural blood patch showed near total regression of the subdural hematoma in both sides.

DISCUSSION

Anatomical Considerations

The SSS is a large midline structure situated among the dura mater lining inner table of the calvarium superiorly, leaves of the FC laterally and along attached margin of the FC inferiorly. It

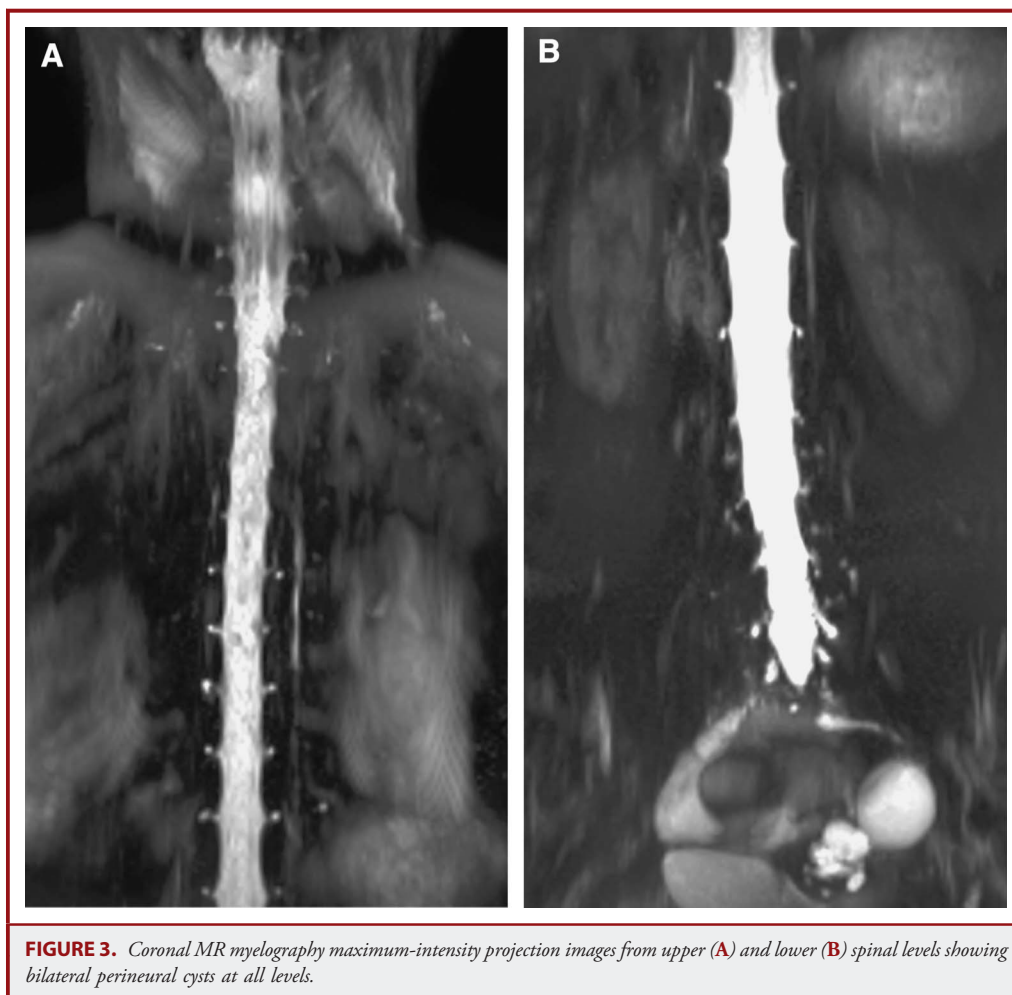
starts at the foramen cecum and goes toward the internal occipital protuberance, where it is connected with straight and transverse sinuses to form torcular Herophili. Anatomic variations of the SSS are uncommon. Although segmental abnormalities are more frequent, all segments may be involved in rare occasions.^{1,2} Anatomic variations of the SSS described in the literature are duplication (segmental or complete), hypoplasia (unilateral or complete), and atresia (segmental).²⁻⁵ However, total or segmental agenesis of the SSS has not been described previously.

The FC is a sickle-shaped, vertically situated, midline structure formed by folds of the dura mater in the interhemispheric fissure. It is attached to the calvarium, starting from the crista galli of the ethmoid bone anteriorly and extending posteriorly to join to the tentorium cerebelli. It contains the SSS at the superior convex margin and the inferior sagittal sinus at the inferior concave margin. Anatomic variations of the FC are so rare that there is very little information in the literature. The variations described in the literature are fenestration, hypoplasia, and partial agenesis.^{6,7} However, total agenesis of the FC has not been reported previously like that of the SSS.

Embryological Considerations

The FC is differentiated from the interhemispheric mesenchyme and develops in 2 parts at different times: the anterior and posterior parts. The anterior part differentiates somewhat earlier than the posterior part that develops in association with the tentorium cerebelli. Eventually, all parts of the falx join each other to create a single midline structure.⁷

The interhemispheric mesenchyme that will become differentiated to the FC contains a plexus of veins on the dorsal surface



of the primitive telencephalon called the sagittal plexus.⁸⁻¹⁰ The sagittal plexus is continuous with the tentorial plexus that lies within the mesenchyme that will form the tentorium cerebelli, forming the sagittotentorial plexus.⁸⁻¹¹ The SSS develops from the sagittal plexus and in 2 parts like the FC.¹¹ At the first 8 weeks of embryological development, the anterior part of the sagittal sinus differentiates from a longitudinal venous channel that is formed by the sagittal plexus and has a pattern similar to that seen in adults. During this stage, developing cerebral hemispheres become separated by interhemispheric mesenchyme forming the FC.⁸ Unlike the anterior part, the posterior part of the SSS develops through elongation during displacement of the tentorium cerebelli inferiorly and the cerebral hemispheres posteriorly. By 11 weeks, the SSS becomes positioned similar to the adult pattern with a well-defined shape.^{9,10} The FC attaches to the developing calvarium around this stage.

In this regard, because they share an anatomical and embryological origin, cerebral falx abnormalities may be related to SSS abnormalities and vice versa.

Imaging Findings and Related Consequences

In imaging studies including CT, MRI, and DSA, total agenesis of the SSS is primarily characterized by the absence in appearance or lack of contrast filling in all segments of the SSS. Additionally, absence or other anatomical variations of such a large dural venous sinus might affect the intracranial venous drainage pathways in different ways, especially leading to the development of variant drainage patterns to compensate drainage of the brain structures properly.^{2-5,12} Venous drainage affected on account of the absence of the SSS can be compensated by some other enlarged venous structures situated around the region of the SSS. In our case, those are the tentorial sinus, frontal hemispheric veins, and parasagittal large venous channels. In clinical practice, these enlarged venous structures can lead to a misdiagnosis as arteriovenous fistula or arteriovenous malformation.

Imaging findings of agenesis of the FC in MRI and CT include primarily total absence of anterior and posterior parts of the FC at the interhemispheric fissure. On rare occasions, this abnormality may cause diagnostic confusion regarding subdural pathologies.

For instance, subdural hematomas can cross the midline, despite the fact that subdural hematomas are known to continue along the FC but cannot cross the midline because of the meningeal reflections made by falx. Moreover, some secondary abnormalities in appearance might be seen such as cerebral gyral interdigitation (gyri crossing the midline), which may also be seen associated with Chiari II malformation owing to hypoplasia and/or fenestration of the FC.¹³ In our case, findings of total agenesis of the FC were considerable posterior extension of the confluent nonrestricted subdural hematoma and gyral interdigitation, as well as the lack of the FC at the interhemispheric fissure.

CONCLUSION

Agenesis of the SSS and FC are extremely rare variations. Agenesis of the SSS results in the development of alternative venous pathways and may lead to misdiagnosis as dural arteriovenous fistulas. FC agenesis may cause diagnostic confusion because subdural pathologies such as hematomas can cross the midline in rare occasions.

Disclosure

The authors have no personal financial or institutional interest in any of the drugs, materials, or devices described in this article.

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COMMENTS

The authors submit a revision to their manuscript entitled "Total Agenesis of Superior Sagittal Sinus and Falx Cerebri in a Patient Who Has a Subacute Subdural Hematoma Crossing Midline: Case Report" and describe the unusual case of a 49-year-old woman presenting with bilateral subdural hematomas due to intracranial hypotension. Extensive imaging workup revealed an extremely rare and previously unreported anatomic variation with total agenesis of the superior sagittal sinus and the falx cerebri. This is a fascinating case, but it should be noted that this is an anecdotal report and limited conclusions can be drawn from this. Based on the information available to us, the authors are unable to place the case presented here within the context of known developmental variants and anomalies and merely report on the finding. This is disappointing overall, but the reporting of this entity may enable readers of this report to be able to use this information and connect the dots some day.

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The authors report an extremely rare but fascinating case of congenital absence of the superior sagittal sinus (SSS) and falx cerebri (FC) accompanied by multiple spinal perineural cysts in a 49-year-old woman with headaches and a continuous, bilateral, septated subdural hematoma. The three congenital anomalies are thoroughly documented with excellent imaging studies. This is probably the first reported case of agenesis of the SSS and FC. The authors provide an excellent review of the known anomalies of the SSS and FC, as well as of the embryology of these structures. The SSS and FC both originate from the interhemispheric mesenchyme, so it makes sense that agenesis of their primordium would result in absence of these two structures. The link between the absent SSS/FC and the multiple spinal perineural cysts is more difficult to understand and will present an interesting conceptual challenge to central nervous system embryologists.

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