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Hypothetical Mechanism of the Formation of Dural Arteriovenous Fistula – The Role and Course of Thrombosis of Emissary Vein and Sinuses

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1. Introduction

Dural arteriovenous fistula (DAVF) is the acquired and progressive arteriovenous (AV) shunt disease on or between the dura matter, and its etiology is still controversial¹⁻⁴. This disorder occurs not in the whole dura but at very specific locations. DAVF can be divided into two types based on the intervention of the drainage route and affected sinus; sinus type and non-sinus type. The sinus type has the shunt at the sinus wall or dural vein, and includes DAVF at the cavernous sinus (CS), transverse-sigmoid sinus (TS-SS), anterior condylor confluence (ACC), and superior sagittal sinus (SSS). The non-sinus DAVF has the shunt on the dura and directly drains into the pial veins, and includes tentorial, ethmoidal, craniocervical and spinal DAVF. However, even the sinus type DAVF ultimately changes to the isolated sinus with cortical reflux due to progressive sinus occlusion, similar to the non-sinus type. Such seemingly separated and complex pathogeneses of DAVF remain elusive.

2. Previous theory about DAVF etiology

Previous recognition of the etiology of DAVF has been directed to sinus hypertension^{1,4} and thrombosis^{5, 8}. It is true that such abnormal situations may create the experimental AV shunt^{1,4}. However, if one considers that the sinus hypertension is the initial trigger, it should be caused secondary to the thrombosis or outlet stenosis. It is unreasonable to adopt this theory into non-sinus type, because this type has no correspondence with the sinus. In other hand AV shunt formation can easily create the condition of sinus hypertension. Thus, the conventional discussion over the etiology of AV shunt formation between sinus occlusion and sinus hypertension is just a chicken-or-egg question. We consider that sinus hypertension concerning sinus wall hypertrophy may not be the cause, but rather one factor in the development of DAVF. Our theory based on inflammatory initiation affecting EV can explain both types of DAVF and subsequent development with pathological changes of the drainage route is not contradictory to the previous sinus-oriented theory.

3. Hypothesis

Etiologically, DAVF is revealed secondary to causes such as trauma, inflammation, or sinus thrombosis⁵⁻⁸. However, most causes are idiopathic and independent of the preceding hematological and immunological impairments. Therefore, comprehensive factors concerning the initiation of DAVF that covers all of the pathological features should be considered. From this perspective, we focused on the location of emissary veins (EV) and discovered that the distribution of EV definitely corresponds to those of the location of DAVF (Fig. 1). According to this previous consensus and to new information, we propose following hypothesis concerning the development of DAVF which focuses on the emissary vein.

A Postero-anterior view
B Lateral view 1. parietal (parasagittal) emissary vein (EV), 2. occipital (torcular) EV, 3. mastoid EV, 4. condylar (condyloid) EV, 5. hypoglossal EV, 6. frontal (ethmoidal) emissary vein (EV), 7. petrosquamosal EV, 8. foramen ovale EV, 9. foramen spinosum EV, 10. foramen lacerum EV, 11. Basilar plexus EV and Trolard's inferior petrooccipital vein (venous plexus of Raktzik)

Fig. 1. Distribution of emissary veins

Emissary veins connecting the intracranial and extracranial venous system through the bone are distributed in specific parts of the vault or base of the skull⁹. They are usually accompanied by and penetrate together through the same foramen with emissary arteries (transosseous perforating arteries) (Fig. 2A). Fig. 2 shows the scheme of this process in the DAVF at SSS (a representative of sinus type DAVF). First, some inflammatory reaction occurs at the penetrating site of EV. It may be reasonable to consider the cause of inflammation to the infection of adjacent tissue such as sinusitis and mechanical inflammation after trauma (including catheter intervention). Sinus thrombosis is occasionally observed before the occurrence of DAVF⁷, however, such thrombosis might be the result of focal inflammation. In most cases with DAVF, inflammation will develop undetected or as an autoimmune allergic reaction. Local inflammation may expand with expression of various cytokines, cause vessel dilatation, and open the physiological AV shunt at the level of capillary vessels (Fig. 2B).

A Normal site. Emissary vein (EV) and artery (a) is penetrating through a foramen of the parasagittal skull. EV is connected with the venous lacunae (b). Meningeal arteries (c) have no connection with SSS (d) and cortical vein (e).

B Neovascularization (arrow) and vessel dilatation induced by dural inflammation

C AV shunt formation at the level of dural arteriole and penetration into the sinus (initial stage of DAVF). Note the shunt flow draining into the sinus as well as EV (double arrow).

D. Shunt development with thrombosis of an emissary vein (asterisk) and recruitment of distal arteries from anterior falx artery (f) and posterior meningeal arteries (g).

E. Maturation of DAVF with the reflux to cortical veins (red arrow) due to sinus occlusion (white arrow). Note the further recruitment of feeders from the other side or transosseous branches (h).

Fig. 2. Mechanism of development of the DAVF at the superior sagittal sinus (SSS) as a representative of the sinus type DAVF.

In sinus type DAVF, a micro AV shunt between the emissary artery and vein will enlarge to the adjacent sinus wall. The increase in shunt flow triggers drainage into the sinus, and subsequently changes the main drainage route from the EV to the sinus. As a result, the sinus will be occupied by the shunt flow more than the normal intracranial venous outflow pathway (Fig. 2C). While this shift in shunt flow direction decreases the role of EV as the drainage pathway, the developed and swollen emissary artery compresses the accompanied EV and impairs the drainage flow. This results in occlusion of the EV (Fig. 2D).

This degeneration of the EV is an important process in the formation of a well-known style of sinus type of DAVF. However, in some cases of DAVF at CS and SSS the EV may remain patent and serve as a drainage route to the pterygoid plexus or parietal vault. Also an EV that connects with diploic veins can form an enlarged intraosseous venous lake, and appear as a new channel of sinus or duplication. The shunt recruits other dural feeders from the distant and contralateral parts due to angiogenetic and hemodynamic factors, and forms the extended vascular network as a new DAVF₁₀. Extracranial arteries on the skull or under the skull base are often mobilized through the bone. Such active recruitment of feeders is considered to be due to angiogenesis enhanced by the expression of vasculogenetic factors at the affected dura (including vascular endothelial growth factor (VEGF))¹¹⁻¹⁶.

The next key process in maturation of the DAVF is the occlusive change of the draining system. Although draining pathway may finally occlude due to intrasinus thrombosis associated with hypercoagulopathy, the essence of the occlusive mechanism should be hypertrophy of the sinus wall₁₂. This occlusive process is typical in the DAVF at CS₁₇. Its drainage route gradually occludes from the inferior petrosal sinus and superior ophthalmic vein₁₁, and occasionally causes a paradoxical worsening of visual acuity and chemosis with ocular hypertension following occlusion of the anterior drainage route. During this progression, the thrombophilic abnormalities characteristic of DAVF are also reported₁₈₋₂₂. In some cases such thrombotic change of drainage route may occur in the initial stage preceding the development of the shunt.

Next, the same process progresses in the upstream side because the remaining upstream drainage with more hemodynamic stress may yield the hypertrophic change of sinus wall_{12, 23}. As a result, the meeting point of the shunt flow will be isolated, and shunt flow without exit to the sinus may reflux into cortical veins (Fig. 2E). Such a matured and aggressive type of DAVF with an affected isolated sinus or dural vein₂₄ (Fig. 3), may be the final expression

A. DAVF at cavernous sinus, B. DAVF at transvers-sigmoid sinus

Fig. 3. Examples of matured sinus type DAVF

of this process. However, this final isolated part of the DAVF will not be always consistent with the first trigger point of a micro AV shunt, because the inflammatory extension and recruitment of many dural arteries will easily cause the movement of the main shunt point. It is quite easy to adopt this mechanistic hypothesis to non-sinus type DAVF. Fig. 4 demonstrates the scheme of the development of ethmoidal (anterior skull base) DAVF as a representative of non-sinus type DAVF.

A. Normal state. a; frontal cortex, b; frontal emissary vein, c; dural branches from anterior ethmoidal artery
B. Neovascularization and dilatation induced with dural inflammation (#)
C. AV shunt formation at the level of dural arteriole and reflux into an EV (initial stage of DAVF)
D. Shunt development and increased reflux into the varicose cortical vein (white arrow) following the occlusion of an emissary vein

Fig. 4. Mechanism of development of the DAVF: an ethmoidal (anterior skull base) DAVF as a representative of the non-sinus type DAVF.

EV at the anterior skull base (Fig. 4A) connecting with the cortical vein will create, secondary to the ethmoidal inflammation, a micro AV shunt at the skull base dura (Fig. 4B). Subsequently the EV will occlude according to the same process as described above (Fig. 4C, D). As a result, all the shunt flow supplied from ethmoidal arteries drains into the cortical veins, which is the common style encountered in the clinical setting. The pathological process at the spine or craniocervical junction DAVF can be explained by the same mechanism.

4. Atypical DAVF

1. **Tentorial DAVF** It is true that there is no EV on the tent. The name of this type of DAVF may come from the participation of tentorial artery, however the main shunt point is not on the tent but at the clival dura (Fig. 5). The tentorial artery as the feeder usually creates the AV shunt just posterior to its origin, and the shunt flow immediately drains into the subtentorial venous complex. Although there are some classifications of various type of tentorial DAVF²⁵, the most common type of tentorial DAVF at the clivus may be caused by the pathological process of a concerned EV, petrosal bridging vein²⁶, that connects between the ventral venous system in the posterior fossa and the basilar plexus. The exceptional tentorial DAVFs affecting the posterior tentorial sinus or the confluence are considered to be variants of TS-DAVF with the influence of EV at the petrous and occipital skull.

Fig. 5. Tentorial DAVF. The lateral view of the left carotid angiogram showing the dilated feeder from the tentorial artery (arrow) and AV shunt at the clival portion (asterisk) immediately draining into the preponine venous network.

2. **DAVF at the anterior condylor confluence** DAVF at the anterior condylor confluence has been newly defined as the generic clinical entity²⁴, and includes DAVF of the inferior petrosal sinus, DAVF of the marginal sinus, hypoglossal DAVF, DAVF of the anterior condylor vein within the hypoglossal canal, and jugular foramen DAVF²⁷. Anterior condylor confluence (ACC) is extracranially located, and is a major venous crossroad at the posterior base of the skull. Tributaries from the hypoglossal canal, petroclival fissure, and the vertebral venous plexus meet together at the ACC and drain into the jugular bulb

through multiple channels²⁸. As seen in previous nomenclature, one of the important drainage routes is the anterior condylor vein is the EV passing though the hypoglossal canal. However, in most cases, the anterior condylor vein has been already occluded, and other venous systems (including the lateral condylor vein, inferior petroclival vein, and inferior petrosal sinus) may function as a drainage route via ACC. Specific characteristics of this type of DAVF include patients suffering from strong tinnitus just when the DAVF is initially formed. Hypoglossal palsy develops in some cases. DAVF at the ACC tends to be diagnosed in the early stages. Therefore, as the original drainage route, the anterior condylor vein occasionally remains one of the draining veins.

3. Vault DAVF This rare type of DAVF is located at the temporal or occipital convexity, and is a non-sinus type DAVF. An aggressive feature of this type of DAVF is that it directly drains into cerebral cortical veins. According to our theory, it may be caused by the focal inflammation around the atypically located EV, or due to the congenital focal connection between pial and dural veins.
4. Multiple, de novo, recurrent DAVF. These clinical features cannot be explained with the single inflammation theory, and spreading or multifocal inflammation should be considered (Fig. 6). Although recurrence of the same lesion can be due to incomplete occlusion of the shunt²⁹, de novo creation of DAVF independent from the previous ones may follow the newly developing process, possibly be promoted with constitutional factors.

Fig. 6. Multiple DAVF Multiple DAVF at superior sagittal, transverse and sigmoid sinus (arrows)

5. Supporting clinical situation

This hypothesis is supported by some familiar features encountered in clinical cases. First, in the case of mature SSS-DAVF, shunt flow usually drains into the cortical vein through an isolated sinus with the particular congestion of pial veins. However, in spite of such an aggressive type with reflux to the cortical vein, SSS is still patent in some particular cases. This unusual situation suggests the influence of EV at the initial location of the micro AV shunt. As seen in Fig. 7, the parasagittal (parietal) EV has no direct connection with SSS itself and drains from venous lacunae. The abnormal state mentioned above can be interpreted as

follows; the occlusive change of drainage site might occur at the channel between venous lacunae and SSS after the formation of AV shunt, therefore SSS as the normal cortical drainage route is independent from DAVF and can be preserved. It may suggest that the shunt point is located not the sinus wall of SSS but venous lacunae, exit of EV. In the early stage of CS-DAVF without ocular symptoms there are various drainage routes into the pterygoid plexus as well as the superior ophthalmic vein and inferior petrosal sinus. Similarly, the anterior condylor vein is patent in the initial stage of ACC-DAVF. In such young DAVF, EV of the foramen ovale or foramen lacerum and hypoglossal EV still remain as the original drainage pathway. This fact suggests the possibility that the EV plays an important role in the initial stages of newly developed DAVF.

One often encounters a multiplicity about the location of TS-SS-DAVF. This fact is also explainable using the present hypothesis. At the confluence, the lateral side and the sigmoid junction, the initially affected EVs: may be torcular, petrosquamosal and mastoid EVs, respectively.

Unfortunately, our hypothesis has not yet been proven in a pathological specimen of clinical cases as well as from the animal experiments. Further, it is difficult to explain the etiology of DAVF in locations without emissary veins or those without arterial supply coming from emissary arteries with the exception of osteodural AV shunts. Although our theory was not based on the anatomical, physiopathological or clinical observational convincing background, if the very early stage of DAVF is incidentally found, inflammatory investigation into the inflammation and meticulous observation of the flow change will be helpful to predict the development of DAVF and also to support this theory.

6. Conclusion

According to previous theories concerning the mechanism of DAVF, it is very important to consider the occlusive pathway and hypertension of the affected sinus. However, previous theories did not indicate an initiation of this pathological situation which can explain both sinus and non-sinus type DAVF and the promoting factors to develop the extension of DAVF. We propose a new theory: the inflammatory vascular network at the penetration site of emissary veins may induce local shunt formation. Subsequent occlusion of the affected EV completes the usual figure of DAVF, and, the sinus (venous) occlusive pathway and arterial recruitment are important steps in the maturation of the DAVF. Previous mechanistic hypotheses focusing on sinus hypertension and sinus thromboses cannot explain the pathogenesis of non-sinus type of DAVF. Although the etiology of DAVF may be concerned by the thrombo-occlusive change of sinus, and our theory is only a speculation without the base of experimental study, it may enable to understanding the common etiology of the two (sinus & non-sinus) types of DAVF, and is not contradictory to the previous sinus-oriented theory. Pathological proof of the initial stage of DAVF will be mandatory.

Abbreviations: DAVF: dural arteriovenous fistula, EV: emissary vein, CS: cavernous sinus, SSS: superior sagittal sinus, TS: transvers sinus, SS: sigmoid sinus, ACC: anterior condylor confluence, CCJ: craniocervical junction

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