Primary empty sella in association with superior sagittal sinus thrombosis and dural arteriovenous malformation

Case report

TOSHIKIC HAI SA, M.D., SHINICHI YOSHIDA, M.D., TOSHIYUKI OHKUBO, M.D., KOHIKI YOSHIKAWA, M.D., AND TOHRU MACHIDA, M.D.

Departments of Neurosurgery and Radiology, Kanto Rosai Hospital, Kawasaki; and Department of Radiology, University of Tokyo Faculty of Medicine, Tokyo, Japan

A 61-year-old woman under treatment with oral medroxyprogesterone acetate following surgery for breast cancer developed diplopia and papilledema due to raised intracranial pressure (ICP). Radiological studies disclosed an empty sella turcica, a dural arteriovenous malformation (AVM), and superior sagittal sinus thrombosis. The sinus thrombosis was considered to have been a side effect of the medroxyprogesterone acetate medication. The association between a primary empty sella and raised ICP is briefly discussed, along with the cause-and-effect relationship between a dural AVM and venous sinus thrombosis.

**KEY WORDS** • empty sella • dural arteriovenous malformation • sinus thrombosis • medroxyprogesterone acetate • intracranial pressure

Primary empty sella can be defined as the condition where the sella turcica is filled with cerebrospinal fluid (CSF) owing to herniation of the subarachnoid space through the incompetent diaphragma sellae, without prior surgery or irradiation for an intrasellar tumor. It is of interest that primary empty sella is occasionally seen in association with raised intracranial pressure (ICP). A number of cases of dural arteriovenous malformation (AVM) associated with venous sinus thrombosis have been documented. The cause-and-effect relationship of these disorders is still a controversial issue.

We report the case of a patient with primary empty sella in association with a dural AVM and superior sagittal sinus (SSS) thrombosis; the presenting symptoms were diplopia and papilledema due to raised ICP, which was considered to have been caused by both a dural AVM and SSS thrombosis. The association of a primary empty sella with raised ICP, along with the relationship between a dural AVM and sinus thrombosis, is briefly discussed.

**Case Report**

This 61-year-old woman with a 2-week history of diplopia was referred to our hospital. She denied headaches or decrease of vision. She had been treated with medroxyprogesterone acetate (200 mg three times a day) for 17 months, following surgery for breast cancer. Examination. Her height was 154 cm, weight 70 kg, and blood pressure 146/90 mm Hg. Neurological examination demonstrated a sixth cranial nerve palsy, complete on the left and partial on the right, with bilateral papilledema. Visual acuity was 1.0 in the right eye and 1.2 in the left eye. Visual field testing revealed enlarged blind spots bilaterally.

There was no laboratory evidence of hypopituitarism. Lumbar puncture with the patient in the lateral recumbent position showed an opening pressure of 295 mm H$_2$O. The CSF protein and sugar content were within normal limits. Skull x-ray films showed the sella to be of normal size. On computerized tomography (CT), the content of the sella was of low density. Mag-
Empty sella with sinus thrombosis and dural AVM

Fig. 1. Left: Magnetic resonance T₁-weighted image showing the flattened pituitary gland and the sella filled with cerebrospinal fluid. Right: Metrizamide computerized tomography cisternogram showing the sella filled with contrast medium.

Magnetic resonance (MR) images revealed that the sella was filled with fluid of the same intensity as CSF; the flattened pituitary gland was also visualized (Fig. 1 left). On metrizamide CT cisternograms, the sella filled with contrast medium (Fig. 1 right). Cerebral angiograms (Fig. 2) disclosed a dural AVM involving the SSS and no opacification of the middle portion of the sinus. The AVM was fed bilaterally by the superficial temporal arteries and middle meningeal arteries. The patient was diagnosed as having a primary empty sella, a dural AVM, and SSS thrombosis. Her course of medroxyprogesterone acetate, which was incriminated as the cause of the SSS thrombosis, was discontinued.

Operation. Transcatheter embolization was performed with polyvinyl alcohol and tiny pieces of silk. External carotid angiography after the embolization procedure demonstrated disappearance of the dural AVM.

Postoperative Course. The patient’s sixth cranial nerve palsy rapidly improved, and ophthalmological examination showed that the papilledema had almost disappeared, although at lumbar puncture the opening pressure was still 220 mm H₂O. On discharge, her external ocular movements were full and smooth, without papilledema, but she noted slight diplopia on left lateral gaze. Follow-up angiography performed 6 months after discharge revealed no reappearance of the dural AVM. The middle portion of the SSS was not opacified as before. At that time, she did not complain of diplopia.

Discussion

Primary Empty Sella and Raised ICP

The description of the “empty sella” dates back to 1951, when Busch⁵ introduced this term to document the appearance at necropsy of the sella turcica when the diaphragma sellae was incomplete or formed only a small peripheral rim. The author examined the sellae of 788 patients with no known pituitary disease; he noted that only 42% had a complete diaphragm and that in 5.5% the diaphragma sellae was a peripheral rim of tissue 2 mm or less with the pituitary gland flattened to the bottom of the sella.

Fig. 2. Upper: Right internal carotid angiogram, lateral view, showing occlusion of the middle portion of the superior sagittal sinus. Lower Left: Right external carotid angiogram, anteroposterior view, showing the right middle meningeal artery feeding the dural arteriovenous malformation (AVM). Lower Right: Left external carotid angiogram, anteroposterior view, showing the left superficial temporal and middle meningeal arteries feeding the dural AVM.

In 1962, Colby and Kearns⁶ described a patient who developed visual impairment after a transient improvement in vision following irradiation to an intrasellar tumor. The operation performed with the presumption of its recurrence disclosed that the sella contained only some scar tissue and the optic apparatus instead of a recurrent tumor; this complication of irradiation was originally referred to as the “empty sella syndrome.” Later this term came to be used in a wider sense for cases where the sella was filled with air at pneumoencephalography. Subsequently, Weiss and Raskind⁷ emphasized the importance of distinguishing between two types of empty sella; the “primary empty sella” refers to that without prior surgery or irradiation for an intrasellar tumor, while the “secondary empty sella” refers to that related to such procedures. Now, the di-
agnosis of an empty sella can be made by CT cisternography or MR imaging. Interestingly, clinical data show that the majority of patients are middle-aged, multiparous, and obese women.

How the primary empty sella develops is still unclear, but the presence of an incomplete or deficient diaphragmatic sellae is generally considered to be a principal causal factor. One of the potential mechanisms by which subarachnoid cisterns extend into the sella occurs when chronically or intermittently raised ICP is transmitted through an incomplete or deficient diaphragmatic sellae. Although raised ICP is not a sine qua non for its formation, many cases of empty sella associated with raised ICP have been documented. In previously reported cases, the causes of raised ICP have been intracranial tumors or hydrocephalus, while patients without such a condition have been diagnosed as having pseudotumor cerebri or benign intracranial hypertension. Our case is unique in that sinus thrombosis and a dural AVM, both of which have the potential to elevate ICP, were the cause of raised ICP.

Dural AVM and Venous Sinus Thrombosis

Coexistence of a dural AVM and sinus thrombosis is not uncommon. Although occlusion of the SSS is rare in comparison with that of the cavernous or transverse sinus, eight cases of dural AVM associated with SSS thrombosis have been reported. With regard to the relationship between a dural AVM and sinus thrombosis, two different speculations have been presented; one is that dural AVM develops following the organization and revascularization of thrombosed sinus, and the other is that sinus thrombosis is secondary to the venous endothelial damage resulting from high and turbulent blood caused by dural AVM.

Houser, et al., documenting two cases in which dural AVM's developed following dural sinus thrombosis, suggested that dural AVM's of the transverse sinus were acquired lesions. Their postulation was that indigenous dysplastic dural vessels present within the sinus develop and establish a direct artery-to-sinus communication during the organization of an intrasinus thrombosis; thus, dural AVM's grow as a form of revascularization and neovascularization. Chaudhary, et al., reported two similar cases, and suggested that the pathogenesis of dural AVM's might be "growth" of the dural arteries normally present in the walls of the sinuses during the organization of an intraluminal thrombus.

On the other hand, Handa, et al., reported a case of dural AVM associated with occlusion of the sigmoid sinuses, suggesting that turbulence of the blood flow might be caused by direct arteriovenous shunt of a large amount of blood and hence the nearby veins were mechanically damaged. Seeger, et al., radiographically observed progression of thrombus in patients with thrombus formation in the cavernous sinus and carotid-cavernous fistula. They hypothesized that the arterial shunt into the venous sinus elevated venous pressure to a critical point at which the flow in the tributaries is decreased, resulting in blood stasis within the sinus.

In our case, the dural AVM probably developed secondary to the SSS thrombosis, since it is most conceivable that medroxyprogesterone acetate, which is known to complicate thromboembolic diseases, produced the SSS thrombosis. In regard to the treatment, we chose transcatheter selective embolization of the dural AVM in the hope that venous outflow and ICP would improve after embolization and that the occluded SSS might in time recanalize following the discontinuance of medroxyprogesterone acetate. Surgical embolectomy would have been a very interesting alternative option, which could have allowed us to observe what would happen to the dural AVM; however, we did not choose this challenge because direct surgery might be harmful and risky to an ICP-elevated brain and we considered that the principal purpose of treatment was to ameliorate the patient's symptoms by lowering ICP. In the event that the symptoms related to raised ICP reappear without recurrence of the dural AVM, ventriculoperitoneal or lumboperitoneal shunting may be necessary and beneficial. Finally, we would like to emphasize the importance and necessity of performing thorough diagnostic procedures, including cerebral angiography, in patients with symptoms of raised ICP.

References

Empty sella with sinus thrombosis and dural AVM


Manuscript received July 13, 1993.
Accepted in final form September 29, 1993.

Address reprint requests to: Toshihiko Haisa, M.D., Department of Neurosurgery, International Medical Center of Japan, 1–21–1, Toyama, Shinjuku-ku, Tokyo 162, Japan.