Usefulness of Multidetector 3D-CT Angiography in the Evaluation of Infantile Perimedullary Spinal Arteriovenous Fistula

Y. IIZUKA, K. SHIMOJI, S. KYOGOKU, T. MAEHARA, Y. YAMASHIRO*

Department of Radiology, *Pediatrics, Juntendo University School of Medicine; Tokyo, Japan

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Summary

We report an infantile huge perimedullary spinal arteriovenous fistula (SAVF) associated with Hereditary-Hemorrhagic-Telangiectasia (HHT), which was treated by glue embolization in one session. Three-dimensional Multidetector Computed Tomography Angiography (3D-MCTA) was useful in pre- and post-endovascular intervention.

Introduction

There has recently been remarkable progress in multidetector 3D-MCTA. Using this method, high-resolution images comparable to those provided by conventional angiography can be obtained following the intravenous administration of only several tens of milliliters of contrast agent. However, few studies have reported the usefulness of this imaging technique in the evaluation of spinal vascular lesions, and only reports concerning the visualization of the artery of Adamkiewicz can be found in the literature¹. In this case report, we present a patient with infantile spinal perimedullary arteriovenous fistula associated with Hereditary-Hemorrhagic-Teleangiectasia (HHT) in which clinically useful pre- and post-interventional information was obtained by 3D-MCTA.

Case Report

The patient was delivered by spontaneous labor on February 25, 1997, in the 40th week of pregnancy. Birth weight was 2.866g. The patient's family history was remarkable for recurrent epistaxis in the father. In January 1999, the child was taken to a local hospital for a fever of 39°C, and a vesicorectal abnormality was observed. Magnetic Resonance Imaging (MRI) and Magnetic Resonance Angiography (MRA) detected a spinal arteriovenous malformation (SAVM).

The child was taken to the outpatient clinic of that institution once a month for urinary drainage and the administration of laxatives before a decision was reached concerning the appropriate definitive therapy. In November 25, 2000, the child was brought to the department of radiology of our hospital to undergo an intravascular therapeutic procedure.

In the 3D-MCTA study by Aquillion (Toshiba Medical, Tokyo, Japan), a 22-gauge needle was placed in a medial vein of the upper arm and 36 ml of contrast medium was injected manually. Scanning was started approximately 30 s later.

The helical pitch selected was 5.5. ALA-TOVIEW V1.31 (Silicon Graphic, U.S.A.) interpreted the image data obtained using multi-

Figure 1 Three dimensional reconstruction arteriography obtained by multidetector helical computed tomography confirm the arteriovenous malformation with the cutting images of the spinal canal.

planer reconstruction (MPR) images and volume-rendering 3D reconstruction images. The 3D-CTA confirmed the overall structure of the SAVM. (figures 1,2). On December 12, 2000, spinal angiography was performed, which revealed that the radiculomedullary artery from the left thyrocervical artery, (figure 3A) left T7 (figure 3B) and T9 (figure 3C) intercostal artery, and right L1 (figure 3D) lumbar artery were the major feeding vessels. The lesion was exclusively vascularized by the anterior spinal circulation.

Only one shunt was noted on the ventral surface of the spinal cord at the level of T10. The draining vein was associated with a dilated varix. Based on these findings, the diagnosis was huge perimedullary SAVF with multiple feed-

ers, a single-opening fistula, and a dilated perimedullary vein with a varix pouch. The presence of multiple cerebral AVMs (figure 6) and a pulmonary AVM in the left upper lobe, segment 3 (figure 7) was also confirmed, and hepatic AVM was ruled out. Despite these abnormalities, the patient was asymptomatic.

On January 18, 2001, the patient was readmitted to undergo intravascular embolization. Neurological examination at the time of admission showed no weakness of the lower limbs and no remarkable neurological findings other than the vesicorectal disorder due to circulatory insufficiency related to the SAVF. On January 23, after informed consent was obtained from the parents, an intravascular procedure was performed under general anesthesia. Superselective catheterization was obtained with a Magic 0.018 STD (Balt Company, Paris, France) and glue embolization was performed with 3 cc of Histoacryl, 0.2 cc of Lipiodol, and 0.8 g of Tantalum powder (figure 3E).

The abnormal shunt from right L1 was completely corrected, and secondary correction of the other shunts from left T7, left T9, and the left thyrocervical artery was achieved. Inter-costal arteriography of left T9 demonstrated posterolateral spinal artery with a basket formation around conus medullaris, and confirmed upward normal anterior spinal circulation.

After the patient was transferred to the intensive care unit following the embolization procedure, the tachycardia due to high-volume shunt flow was reduced to approximately 80 bpm from 120 bpm, although PACs were noted in the ECG. Improvement was observed in the vesicorectal abnormality noted before treatment.

The patient was discharged on February 10, 2001, after manocystometry and cystography were performed at the Department of Pediatrics. Post-interventional 3D-MCTA (figure 4) and post-interventional curved MPR images (figure 5) confirmed complete disappearance of SAVM except embolized glue deposition.

Follow up cerebral MRA performed every six months showed the growth of small cortical nidus. On October 23, 2001, endovascular intervention for the two cortical nidi was conducted under general anesthesia via the left femoral artery. Single hole fistula located at left orbital gyrus was catheterized selectively with a Magic

0.015 CTD (Balt Company, Paris, France) and glue embolization was performed with 1 cc of Histoacryl, 0.8 cc of Lipiodol, and 0.5 g of Tantalum powder.

The glue occluded the fistula, and achieved the proximal portion of draining vein, which resulted in complete occlusion of this lesion. Another lesion, which was located at left calcarine gyrus, was treated in the same fashion, but resulted in partial occlusion. Angioarchitecture of other micro-AVM lesions, which were the transit type nidus located around the left angular gyrus in patient's dominant cerebral hemisphere followed without treatment.

Embolization for pulmonary AVFs is planned on December 7, 2001 in order to avoid septic emboli or stroke.

Discussion

Infantile SAVMs can be roughly classified into two types. When a nidus is formed, the lesion is found in an intramedullary location in most cases. On the other hand, when an arteriovenous fistula is formed, the lesion is usually found in an intradural extramedullary location, with the arteriovenous fistula on the surface of the spine². To our knowledge, meningeal arteriovenous fistula, as seen in adults, has not been seen in infants. According to Berenstein's report, complicated malformations associated with infantile spinal arteriovenous fistulas, such as mucosal dermal lesions, vertebral arteriovenous fistulas, and Cobb syndrome, have been reported in approximately 28% of cases. SAVMs are recognized in 9% of patients with Rendu-Osler-Weber syndrome and in 5% of patients with Klippel-Trenaunay syndrome³. Halbach reported that spinal AVMs were recognized in two of five cases of Rendu-Osler-Weber syndrome and in one of five cases of Cobb syndrome⁴.

The presenting symptoms are related to sudden hemorrhage in most cases. Progressive neurological symptoms are also produced by congestive venous hypertension due to high-flow and high-volume SAVM. On the contrary, in the literature, it has been reported that progressive neuronal shedding may be due to mechanical compression secondary to the circulatory abnormalities surrounding the spinal cord formed by the emissary vein, and a dilated vein was recognized in only 9% of cases. Varix for-

Figure 2 Pre-interventional curved MPR image shows marked dilated vessels of the spinal arteriovenous malformation.

mation and constriction of the emissary vein are frequently recognized, and may be responsible for erosion or deformity of the vertebral arch. Researchers have also reported that such varix formation and constriction of the emissary vein are frequently seen in cases with high-flow shunts at the thoracolumbar level, as was observed in our case⁵⁻⁸.

In the case presented here, the patient has been clinically observed up to about the age of three years without treatment. The optimal timing for the treatment of infantile spinal AVM has not yet been determined, and treatment is associated with the risk of irreversible neurological impairment due to spinal is-

Figure 3 A) Left thyrocervical arteriography demonstrates anterior spinal artery connecting to the radiculo-medullary artery from the left 9 th intercostals artery with a single hole fistula. B) Left 7 th intercostal arteriography shows radiculo-medullary artery connecting to a single hole fistula.at the ventral surface of spinal cord at th elevel Th10. C) Left 9 th intercostal arteriography shows radiculo-medullary artery connecting to a single hole fistula.at the ventral surface of spinal cord at th elevel Th10.

chemia. We plan to closely follow the patient's clinical course.

All four AVM in the cerebral cortex measured 1 cm or less. They were all situated in the cerebral cortex. In reviewing the several case reports of multiple cerebral AVMs, it appears that most of them have been with either so-called Rendu-Osler-Weber syndrome or HHT¹⁰⁻¹².

A few large case series have noted that multiple cerebral AVMs occur in approximately 1% of all cases. Willinsky et al reported 11 cases (6%) of multiple cerebral AVMs in 203 consecutive patients with cerebral AVMs. These

authors attributed this increased preponderance in their series in part to their meticulous and sophisticated angiographic technique with magnification and re-evaluation around endovascular procedures⁹. The occurrence of an exceptional number of multiple cerebral AVMs, which we have arbitrarily defined as three or more lesions appears to be rare. Three concurrent AVMs could be found in six previous literature reports^{9,13-17}, and five separate lesions occurring in a single patient were reported in two papers^{9,14}.

The multiple AVM with H.H.T. posed unique

raphy shows radiculo-medullary artery connecting to a single hole fistula at the ventral surface of spinal cord at the level Th10. D) Left 1st. Lumbar arteriography demonstrates radiculopial artery, which is the biggest feeding artery connecting to a single hole fistula with the dilated draining vein. E) Glue embolization closes the shunt between multifeeders and single-hole fistula.

and difficult management problems. Natural history studies for sporadic AVM have shown that the cumulative annual hemorrhagic risk is approximately 3-4%^{13,18-20}. Current management is based on the natural history of sporadic AVM¹⁰.

Matubara et al reported that hemorrhage from cerebral AVMs is uncommon in patients with HHT, unlike the case for micro-AVMs in adults^{20,21}. Accordingly, it was decided that the patient should be managed conservatively, with careful observation by follow-up MRA and with treatment deferred until the lesions show

signs of expansion or bleeding at the first consultation. But, follow up cerebral MRA showed the growth of cortical AVMs. It is generally agreed that such lesions tend to undergo a gradual pathophysiological and consequent pathologic evolution, which affect the feeding arteries, nidus, and draining veins. The process often produces degenerative weakening within the wall of these vessels, predisposing to rupture. Although the natural history of sporadic cerebral AVM has been well documented, the specific natural course of the cerebral AVM in HHT is not known²³.

Figure 4 Post-interventional three-dimensional reconstruction arteriography shows glue cast and disappearance of the abnormal shunt.

The risk versus benefit of extensive interventions cannot be justified. Two of them were treated by glue embolization to prevent future bleeding. Therapeutic decision-making is usually complex in children with multiple cerebral AVMs and should be approached on a case-by-case basis^{14,24}.

Neurologic complications of HHT, in which the major problems have been attributed to paradoxical emboli through pulmonary AVMs, resulting in either cerebral infarction or brain abscess, have been well known for decades. Other potential mechanisms of cerebral injury related to pulmonary AVMs are systemic hypoxia due to right-to-left arteriovenous shunting, air embolism, and secondary polycythemia. Polycythemia resulting from a pulmonary AVM may predispose to cerebrovascular thrombosis related to a combination of increased blood

Figure 5 Post-interventional curved MPR images confirm complete disappearance of SAVF except embolized glue deposition.

viscosity, large total blood volume, and thrombocytosis.

A previous study has shown a significant reduction of regional cerebral blood flow in patients with polycythemia¹³. Indications for treatment include three broad categories: exercise intolerance, prevention of neurologic complications, and prevention of lung hemorrhage. Serious neurologic events, including TIA, stroke, and brain abscess, occur in 30~40% of patients with pulmonary AVM²⁵. With regard to the pulmonary AVMs in our case, that had been identified previously, the patient was asymptomatic. Pulmonary AVM should be treated to prevent a catastrophic complication by catheter intervention²⁶⁻²⁸.

The descriptions by Rendu, Osler, and Weber stressed the association of cutaneous telangiectasia with epistaxis and their heredi-

tary occurrence, while more recent literature has shown a widespread angiodysplasia associated with this syndrome. This syndrome has various pathophysiological consequences, depending on which organ system is involved. Various vascular malformations have been described in the brain, retina, thyroid, heart, spinal cord, lungs, intestine, liver, spleen, pancreas, kidneys, prostate, cervix, bladder, urethra, diaphragm, vertebrae, major arteries, and aorta²⁹. A recent pathoanatomic study of skin telangiectasia showed that these malformations are actually plexiform arteriovenous fistulas with significant arteriovenous shunting³⁰. In our case, all lesions had presented fistula shunts.

Two distinct types of HHT genetic analysis were identified in 1994. The focus for the first type is on chromosome 9. A mutation in the gene, encoding endoglin, appears to be responsible for this form of HHT. The locus for the second type has been linked to chromosome 12 encoding the activin receptor-like kinase 1. The phenotype of both HHT includes epistaxis and telangiectasia. Patients with HHT type 1 appear to have greater frequency of cerebral and pulmonary involvement. The phenotype for HHT type2 has not been fully characterized as yet, but it appears that the cerebral and pulmonary involvement are less common in patients with HHT type2 than with HHT type1. There are anecdotal suggestions that multiple cerebral AVM may also be more common in families with HHT type1, but no evidence has been published. As there is no widely available DNA-based diagnostic method, careful history-taking and morphological examination will usually provide insight into whether a patient's family has HHT³⁰⁻³².

In the 3D-MCTA study, MPR images were found to be superior for evaluating microvascular structures, the draining vein of the AVM, and the varix. MPR images, which could be reconstructed to match the normal curve of the spine, provided unique diagnostic information that could not be obtained from coronal or sagittal MRI images, since this method permits scanning to be performed in any desired plane. In addition, volume-rendering 3D reconstruction images were useful for understanding the overall structure of the AVM, which perfectly complements the information obtained by conventional angiography.

Figure 6 Magnetic resonance angiography demonstrate asymptomatic multiple cortical small AVM.

Conclusions

A case of infantile perimedullary spinal arteriovenous fistula associated with HHT has been presented. Multidetector 3D-CT angiography was found to be useful for the pre- and post-interventional evaluation of such lesions.

Figure 7 Helical computed tomography show pulmonary AVMs at left segment 3.

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Yuo Iizuka, M.D.
 Department of Radiology
 Juntendo University, School of Medicine,
 2-1-1, Hongo, Bunkyo-ku, Tokyo 113-8421
 e-mail: yuoi@med.juntendo.ac.jp