

Case reports

A case of multiple arteriovenous malformations and diffuse venous abnormalities with facial port-wine stain

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Abstract. A case of left facial port-wine stain with right hemiparesis is reported. Radiologically, two arteriovenous malformations (AVM) and diffuse venous abnormalities were observed in the left hemisphere. AVM were seen in the basal ganglia. One was fed by a perforating artery from the MCA and drained into the superior petrosal sinus, and the other was fed by a perforating artery from the basilar artery and drained through the vein of Rosenthal into an extremely dilated vein of Galen. Venous abnormalities were obstruction and a decrease in the superficial cortical veins, as well as the formation of collateral veins in both the cortex and white matter. This case resembles Sturge-Weber syndrome (SWS), but there were no signs of leptomeningeal angiomas or venous angioma. We have been unable to find any case reports on SWS with AVM or AVM with diffuse venous abnormalities. We discuss the differences between our case and SWS.

Key words: Sturge-Weber syndrome – Arteriovenous malformation – Venous abnormality – Facial port-wine stain.

We examined a case of multiple cerebral arteriovenous malformations (AVM) and diffuse venous abnormalities. Facial port-wine stain (nevus flammeus) was seen on the left side, and paresis of the extremities was seen on the right side. This case could not be diagnosed as Sturge-Weber syndrome (SWS) because the typical leptomeningeal angiomas that should be found by CT or angiography was not present. Some cases of multiple AVM have been reported, but we could not find any case report of AVM with diffuse venous abnormalities and facial nevus, as in our case. This case would appear to be both extremely rare and interesting in relation to SWS.

Case report

After an uncomplicated 41-week gestation, the baby girl was born; the delivery was uneventful. Her birth weight was 4,300 g

(+2.5 SD) and her height 51 cm (+1.0 SD). Her parents and sister were in good health. There was no family history of neurocutaneous disease. The facial nevus was noted at birth. Her psychomotor development was normal and she started to walk without support at 11 months of age. From 12 months of age on, she frequently fell down towards the right side. At the age of 23 months she was referred to Tsukuba University Hospital for evaluation.

On admission, she was a normally developed child with a weight of 11.0 kg (−0.2 SD) and a height of 80.2 cm (−1.2 SD). There was a port-wine stain which occupied the median part of the first trigeminal area and almost the whole area of the left second and third branches (Fig. 1). No other skin lesions were seen. No abnormal findings were observed in the chest or abdomen.

Neurological examination revealed hyperextensibility of the right extremities. Fine movement and muscle tone were decreased in the right extremities. However, the deep tendon reflexes were normal and there were no pathological reflexes. The intraocular tension and fundi were normal. There was no mental retardation. On electroencephalography, relatively low voltage spindles were observed in the left hemisphere.

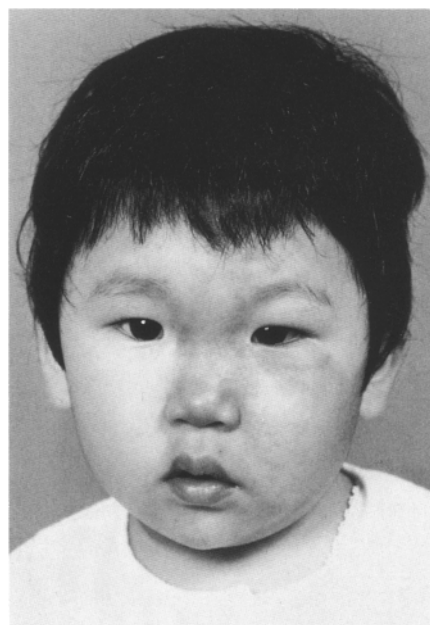


Fig. 1. Port-wine stain seen on the median part of the first trigeminal area and almost the whole area of the left second and third branches

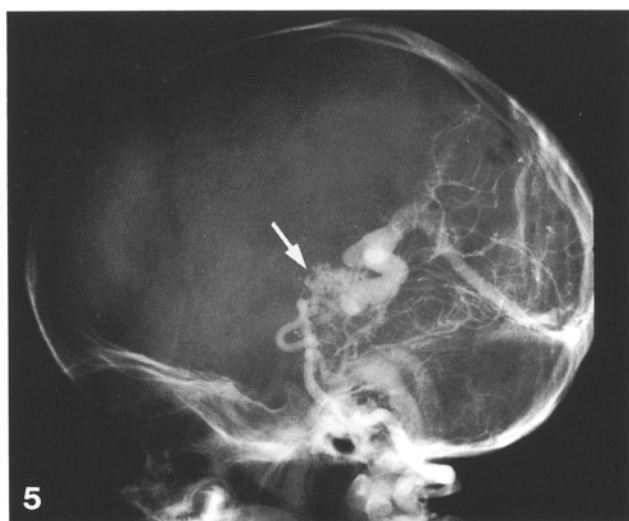
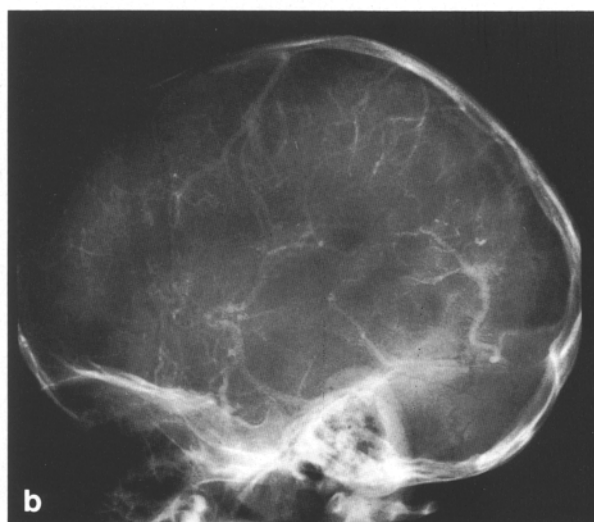
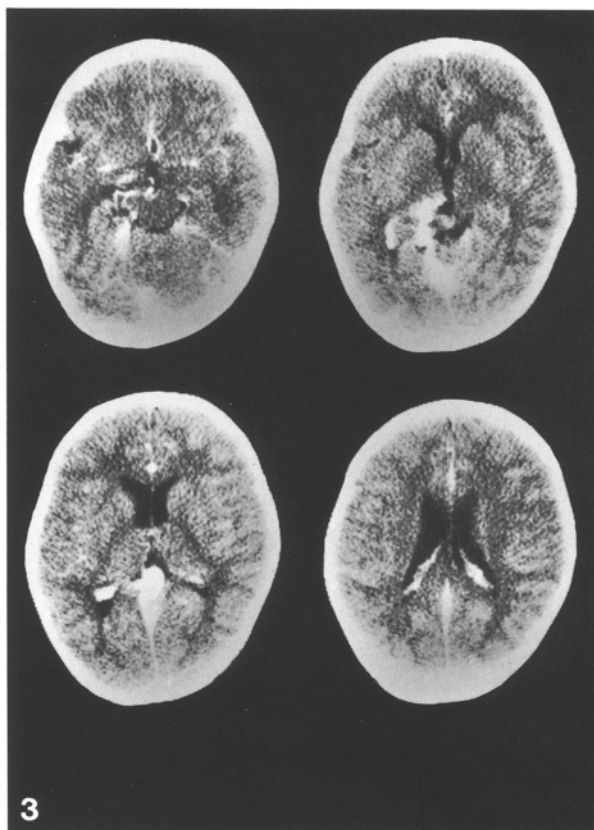
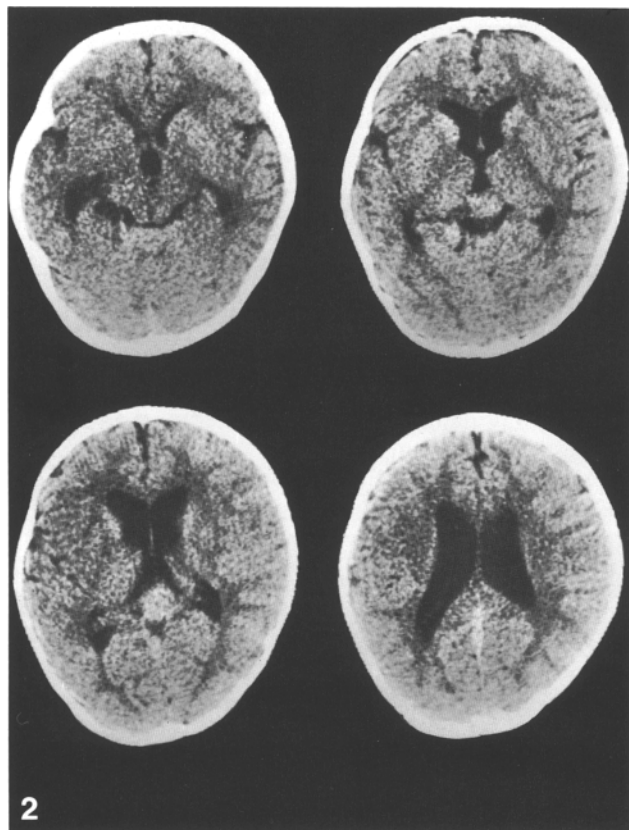


Fig. 2. Plain CT shows dilated lateral ventricles and a tumorlike lesion at the pineal region and no calcification

Fig. 3. Enhanced CT shows that the tumorlike lesion in plain CT was an extremely dilated vein of Galen

Fig. 4. a Left carotid angiography revealed an AVM (↑) on the anterior part of the basal ganglia, which was fed by a perforating artery from the MCA and drained by an abnormal vein (↑↑) to the superior petrosal sinus; **b** decreased cortical veins, formation of collateral veins in both cortex and white matter, and poor filling of the superior sagittal sinus were also shown

Fig. 5. Vertebral angiography revealed an AVM (↑) on the posterior part of the left basal ganglia, which was fed by a perforating artery from the basilar artery, drained off to the vein of Rosenthal and subsequently to the extremely dilated vein of Galen. A patent, primitive, straight sinus was also observed

Table 1. Comparison of our case with SWS, AVM, and with Anderson's case

	SWS	AVM	Anderson's case	Our case
Facial nevus	+	—	+	+
Neurological sign	+	— ~ +	+	+
Multiple AVM	—	— ~ +	—	+
Shunting lesion	—	— ~ +	+	+
at basal ganglia				
Abnormal cortical veins	+	—	+	+
Leptomeningeal	+	—	+	—
angiomatosis				
Venous sinus dilation	— ~ +	— ~ +	+	+
Filling defect	— ~ +	—	+	+
Deep veins				
— Dilation	— ~ +	— ~ +	+	+
— Filling defect	— ~ +	—	—	—
Persistent, primitive straight sinus	—	—	—	+

Radiological findings

There was neither calcification nor other abnormal signs in a plain X-ray film of the skull. Plain CT (Fig. 2) revealed dilated lateral ventricles and a tumorlike lesion at the pineal region without calcification. Enhanced CT (Fig. 3) clearly revealed that this tumorlike lesion was an extremely dilated vein of Galen and that the left choroid plexus was more strongly enhanced than the right.

Left carotid (Fig. 4) and vertebral (Fig. 5) angiography revealed two AVM in the left basal ganglia, decreased cortical veins, developed formation of collateral veins in both cortex and white matter, poor filling of the superior sagittal sinus, and patent, primitive, straight sinus. One of the AVM was situated at the anterior part of the basal ganglia, fed by a perforating artery from the MCA and drained by an abnormal vein to the superior petrosal sinus. Another was situated at the posterior part, fed by a perforating artery from the basilar artery, and drained into the vein of Rosenthal, continuing to the extremely dilated vein of Galen. Some of the veins that should drain to the superior sagittal sinus were interrupted or obstructed. However, right carotid angiography showed no abnormality. The superior sagittal sinus was well filled.

Discussion

The characteristic findings of this case were port-wine stain in the left trigeminal region, paresis on the opposite side, intracranial lesions of the left side that contained multiple AVM in the basal ganglia, obstruction of the superficial veins of the left hemisphere, and formation of collateral veins in the left cortex and white matter. These findings are very similar to those of SWS.

However, it has recently been reported that leptomeningeal angiomatosis is the finding seen mostly in SWS. Maki and Senba [10] analyzed the enhanced CT of eight children with SWS and noted that leptomeningeal angiomatosis appeared to be represented in the area of cerebral surface where calcification will occur in the future.

In reported cases with angiograms of SWS [1, 2, 7, 11, 17], the characteristic SWS findings are mainly abnormal-

ities of the capillaries or small veins, the so-called "capillary blush" or "venous angioma". Larger veins are also affected, e.g., dilation of the deep veins, poor filling of the venous sinuses, and the persistence of primitive veins. Our case had neither an enhanced area in the cerebral surface nor did she have capillary or small vein abnormalities; SWS seems to differ in this respect.

According to an analysis of the literature on AVM series [3–6, 9, 12–15, 19, 20], multiple AVM are found in 4 of 937 cases of AVM (0.43%). This shows that multiple AVM occurs very rarely and, in general, multiple AVM does not accompany any abnormalities of the vein other than dilation of the draining vein [18].

A few cases similar to ours have been reported. Laur [8] reported a case of cerebral AVM with multiple aneurysms, anomaly of the carotid artery and a facial nevus, but made no mention of the venous system. This case had facial angioma and cerebral AVM as in our case, but its abnormalities were mainly arterial. Poser and Taveras [16] classified the vascular abnormalities of SWS and mentioned that Laur's case is typical SWS with AVM. However, we believe his case is atypical because of the absence of obvious venous abnormality. Anderson and Duncan [1] reported a case of SWS with capillary angiomatous malformation in the thalamus and basal ganglia. Their case had venous angioma and was diagnosed as SWS. The coexistence of A-V shunting lesions in the deep cerebrum and diffuse venous abnormality in the cerebral hemisphere (Table 1) resembles the malformations found in our case.

We are unable to ascertain whether diffuse venous abnormality and A-V shunting lesion have the same etiology. However, each abnormality is very rare; hence, the coexistence of two lesions appears to be congenital due to one etiological cause in the prenatal period. We would anticipate that an accumulation of cases similar to ours could explain the etiological mechanisms of these abnormalities.

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