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Surgical Treatment of Patients with Congenital Vascular Malformation-associated Aneurysms

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Abstract *Objectives:* Aneurysms associated with congenital vascular malformation (CVM) comprise critical complication. We review our experience with extracranial CVM-associated aneurysms and attempt to clarify their clinical features.
Patients and methods: The prevalence, site, size and morphology of the accompanying aneurysms of 48 consecutive CVM patients, who were managed at our hospital from 1999 to 2008, were evaluated. After diagnosis or treatment, the patients were followed up, and the recurrence of aneurysms and patient survival were assessed.
Results: CVM-associated aneurysms were found in 14 patients (29%). CVMs were classified according to the Hamburg classification. The patients were classified into groups as follows: four (31%), in the 'predominantly arteriovenous (AV) shunting defect type'; eight (47%), 'combined vascular defects + predominantly AV shunting defects type'; and two (11%), 'combined vascular defects type'. All aneurysms except one situated at the CVM were saccular, whereas nine were fusiform aneurysms; all the ruptured aneurysms and seven out of the nine enlarging aneurysms were saccular. Surgical treatment was performed 8 times in six patients. During the postoperative follow-up period, recurrence and an aneurysm rupture were encountered in one patient each.
Conclusion: Aneurysm is not a rare complication of CVM. It is important to treat CVM before the emergency presents. In addition to the treatment for malformation, regular screening for and proper management of the aneurysms in CVM patients are indispensable.

Congenital vascular malformation (CVM) is a relatively rare disease. It can occur anywhere in the body, including the cranium, face, abdomen or extremities, and can be potentially limb- or life threatening. Congestive heart failure (CHF) could be induced in the presence of arteriovenous (AV) shunts because of increased cardiac output. Major bleeding is also one of the severe complications. Patients also complain of brush or mass lesions, leg-length differences, venous stasis symptoms and cutaneous ulcers.

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The treatment strategy for CVM should focus on both the malformation and the secondary disorders. The goal of CVM treatment, especially for the extra-truncular type originating at the early stage of embryogenesis, should be complete destruction of the nidus that retains the mesenchymal cell characteristics. This includes occlusion of the nidas and fistura or surgery followed by sclerotherapy using ethanol to destroy the nidus and its mesenchymal cells to eliminate the risk of recurrence. Ligation of feeding vessels alone results in the enlargement of numerous smaller feeding and draining vessels.[1] However, aggressive treatment of CVM in the limb always carries a risk of limb loss.

Although a critical complication, CVM-associated aneurysms have infrequently been described.[2] After experiencing a few cases of aneurysm rupture associated with extracranial aneurysm rupture, we reviewed our experience and attempt to clarify its clinical features.

Patients and Methods

We reviewed 48 consecutive patients with extracranial CVM, who were managed at the University of Tokyo Hospital from 1999 to 2008. We diagnosed CVM in the patients on the basis of clinical presentations and/or computed tomography (CT), magnetic resonance imaging (MRI) and/or conventional angiography. CVMs were classified according to the Hamburg classification (Table 1)[3] of vascular anomalies and malformations. Aneurysmal lesions were defined as follows: (1) saccular-form proptosis and (2) dilatation more than twice the normal size. We evaluated the prevalence, site, size and morphology of the aneurysm accompanying the CVM. We then assessed the growth or recurrence of the aneurysm as well as patient survival during the follow-up period; the follow-up visits were conducted in an outpatient clinic once or twice a year.

Results

During the study, 48 patients (24 men and 24 women) with a mean age (SD) of 37.1 (17.9) years at the initial visit were diagnosed with extracranial CVM, that were located in the body trunk in nine patients, in the upper extremity in 14 (right, seven; left, seven) and in the lower extremity in 31 (right, 16; left, 15). Multiple lesions were detected in five patients: in the body trunk and left-lower extremity in two, in the body trunk and right-lower extremity in one patient, in the bilateral lower extremities in one and in the body trunk and both right upper and lower extremities in one. Fourteen patients (29%) had CVM-associated aneurysms. Of these 14 patients, two (14%) exhibited New York Heart Association (NYHA) grade III CHF. On the contrary, 34 patients without aneurysms did not show any signs of CHF. Each CVM was classified according to the Hamburg classification (Table 2). The patients with aneurysms were classified into the following groups: four (31%) patients in the 'predominantly AV shunting defects type'; eight (47%), 'combined vascular defects + predominantly AV shunting defects type'; and two (11%), 'combined vascular defects type'. As per this classification, Parkes–Weber syndrome (PWS) can be included in the 'combined vascular defects + predominantly AV shunting defects type',

Table 1 (A) Hamburg Classification[a,6]-of congenital vascular malformations (CVM) – types; (B) Hamburg Classification of CVM[b]: forms – embryological subtypes.

(A)
Predominantly arterial defects
Predominantly venous defects
Predominantly arteriovenous (AV) shunting defects
Predominantly lymphatic defects
Combined vascular defects
(B)
Extratruncular forms
Infiltrating, diffuse
Limited, localized
Truncular forms
Aplasia or obstruction
Hypoplasia, aplasia, hyperplasia
Stenosis, membrane, congenital spur
Dilation
Localized (aneurysm)
Diffuse (ectasia)

[a] Based on the consensus on CVM through the International Workshop in Hamburg, Germany, 1988, and subsequently modified -Capillary malformation was not included Developmental arrest at the different stages of embryonic life: earlier stage – extratruncular form; later stage – truncular form.
[b] Both forms may exist together; may be combined with other various malformations (e.g. capillary, arterial, AV shunting, venous, haemolymphatic, and/or lymphatic); and/or may exist with haemangioma.

Table 2 Patient characteristics.

Gender (male:female)	24:24
Age (years)	37.1 (range, 2–82)
Mean follow-up period (months)	35.5 (range, 1–146)
Lesion of CVM	
Upper extremity (right:left)	7:7[d]
Lower extremity (right:left)	16:15[d]
Body trunk	9[d]
CVM type (Hamburg classification)	
AV shunting[a]	13 (4 aneurysms, 31% and 1 heart failure)
Combined + AV shunting[b]	17 (8 aneurysms, 47% and 1 heart failure)
Combined type[c]	18 (2 aneurysms, 11% and no heart failure)

[a] Predominantly arteriovenous (AV) shunting defects type in Humburg Classification.
[b] Combined vascular defects + predominantly AV shunting defects type in Humburg Classification.
[c] Combined vascular defects type in Humburg Classification.
[d] Duplicative lesions of the body trunk and left-lower extremity in 2 patients; body trunk and right-lower extremity in 1; bilateral lower extremity in 1; and body trunk, and right-upper and right-lower extremities in 1.