Management of Congenital Vascular Malformation: Overview

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Approaches to patients with congenital vascular malformation (CVM) have been changed not only in the diagnosis but also in the management through the last two decades. Newly established classification and understanding on the anatomic and biologic characteristics of various types of CVM lesions took the leading role to formulate contemporary management principles.

Ultimate goals of the CVM treatment include reduction or extirpation of CVM lesion with minimal recurrence and treatment-related complication rates.

To achieve these goals, minimal invasive or multidisciplinary approach is more frequently adopted in current management of CVM patients.

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In general, following points are usually considered before treatment of CVM patients.

- Treat or "wait and see" – require evaluating risk versus benefits of each side.
- What is an expected natural course of the CVM lesion? – consider expected complications or sequelae when leave it untreated.
- If decided to treat, when is an optimal timing for the treatment?
- What are important structures around the CVM lesion?
- If decided to treat, how can we approach to the CVM lesion without or with minimal damage to the normal tissue or organs around the CVM lesion?
- If decided to treat, how can we reduce rates of recurrence and treatment-related complication?

Not all the CVMs would need the treatment or equally treated because they take different natural course with different prognosis. As described in previous chapters, CVM shows various clinical features and different clinical significance according to the type, anatomic site and extent of the CVM lesion, and age of the patients. For example, arteriovenous malformation (AVM) is considered as a potentially life- or limb-threatening condition in general, while venous malformation (VM) is not.

Head and neck CVMs usually present with cosmetic reasons, and CVM lesion close to the upper airway may cause breathing difficulty.

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Patients with CVM lesions in the extremity often present with pain, swelling, and/or bleeding or lymphatic discharge from skin, while some patients present with serious limb length or size discrepancy, joint contracture, or foot ischemia.

Depth of CVM involvement and anatomic location of the CVM lesion are also important to determine an optimal treatment strategy.

When sclerotherapy is considered for treatment of CVM, chemical skin burn is worrisome in CVM lesion confined to facial skin, while functional defect due to nerve damage is more worrisome for patients with CVM lesion close to the major nerve in the extremity. According to the anatomic location of the CVM lesion, treatment plan can be different considering cosmetic or functional complications related with the treatment.

Age of patient is another important factor in determining treatment strategy for patients with CVM. CVM lesions may present at late days in the life though it is congenital abnormality of blood or lymphatic vessels. Regarding to an optimal timing for the treatment of CVM lesion, early extirpation of CVM lesion is theoretically reasonable. However, endovascular or surgical treatment in pediatric patients is challenging due to small size of blood vessels, limited safety margin of sclerosing agent, and radiation hazard by the endovascular sclerotherapy. Therefore, risk versus benefit analysis should be done before attempting surgical or endovascular treatment of CVM. On an analysis to predict response to the percutaneous ethanol sclerotherapy in patients with venous malformation (VM), we found that no or delayed visualization of drainage vein on an initial direct puncture venogram, well-defined VM margin on MR image, and female gender were predictors of "better response" to the sclerotherapy [1].

Lymphatic malformation showing macrocystic lesion is an exception to the delayed treatment. It can be treated with local percutaneous injection with sclerosing agents.

Sometimes, CVM may be detected at late in life by development of complications of hidden CVM lesion. For example, lower extremity lymphatic malformation (LM) may present with repeated episodes of cellulitis, and pelvic AVM lesion may present with polymenorrhea and chronic anemia. To detect the clinically unapparent CVM lesion, it is required high index of suspicion of CVM. For an optimal management CVM lesion, knowledge in clinical and biologic features of various types of CVM and precise anatomic information are required.

Regarding the optimal treatment method, endovascular embolo-sclerotherapy is most frequently adopted therapy and particularly for patients with surgically inaccessible CVM lesion such as *diffuse infiltrating* type of extratruncular CVM lesions involving extensive regions of tissues. Surgical excision of *limited and localized* CVM lesion can be adopted as long as complete excision of the lesion is available with acceptable risk of complication. Sometimes both treatment methods can be attempted concomitantly or as staged procedures.

However, incomplete or inadvertent surgical excision of CVM lesion can make it worse due to recurrence or even aggravation of the CVM lesion and interruption of endovascular access route.

In this part VI (contemporary management of CVM), authors described various types of treatment modalities for CVM patients according to the types and anatomic location of CVM lesions.

Reference

1. Yun WS, Kim YW, Lee KB, Kim DI, Park KB, Kim KH, Do YS, Lee BB. Predictors of response to percutaneous ethanol sclerotherapy in patients with venous malformations: Analysis of patient self-assessment and imaging. J Vasc Surg. 2009;50:581–9.