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Radical Excision of Low Flow Vascular Malformations-Choice of Treatment in Early Childhood

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Abstract

The low flow vascular malformations are very common in children. The vascular malformations can increase in size and invade locally that could lead to, bleeding deformities and growth defects. There are many modalities of treatment such as embolisation, sclerotherapy and excision. The low flow malformations can be treated and cured with radical excision. This article discuss a few cases treated by radical excision and highlights the importance of surgical treatment in early childhood with the help of microsurgical techniques.

Keywords: Vascular Malformation; Low Flow Malformations; Radical Excision; Sclerotherapy; Embolization.

Introduction

The vascular anomalies are very common in children. They can be either malformations or vasoproliferative lesions. The 'vascular malformations' are always present at birth, enlarge with growth of the child and they never involute in adulthood. They can produce secondary changes like gigantism, local invasion and spread leading to physiological obstruction and deformities. The vascular malformations are subcategorized based upon the histologic tissues like lymphatic, capillary, venous, arterio-venous, and mixed malformations. The low flow malformations are mainly seen in head and neck region and limbs. Generally the low flow anomalies are treated with sclerotherapy and embolisation. The total excision of well-defined low flow lesions is possible and it will give better cosmetic and functional results. In this article, few of such cases treated by surgical method are discussed with literature review.

Materials and Methods

5 children with large vascular malformations treated in a period of 2 years were studied and analysed. The basic investigation of choice was Doppler vascular study and that helps to identify the flow rate and type of vessels and tissues involved. The cases of malformations in the head and neck regions were investigated with CT scan and made it clear about the extend of the malformations. Clinically well-defined masses with low flow rate and small arterial feeders were selected for surgical excision. Even if the lesions appear well defined there were multiple vascular channels extending to the surrounding tissues. A child with large malformation of the neck [Figure 1] had vascular connections to external jugular vein and braches from external carotid artery. The local excision of the lesion was done under microscopic magnification. The limb vascular malformation was done with help of tourniquet control.

Fig. 1: 6 months old child with huge low malformation with attachment to the internal jugular and extension to the parotid.

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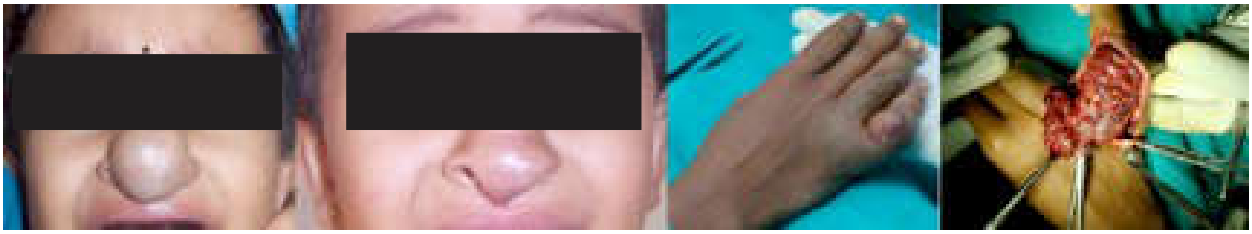

Fig. 2: lip vascular malformation in two children

Fig. 3: Malformations on the nose of a child and hand with early local gigantism

Discussion

The vascular anomalies are a group of conditions with complexity of management due to the varying types of histological components, velocity factors of the blood flow and neoplastic properties of the tissues. There are many modalities of treatment like sclerotherapy, embolisation, excision and even radiotherapy. The complications of treatment are equally significant and hence many of the cases are left alone leading to long-term sequel like bleeding, obstruction to airway, and incurability.

In 1982, Mulliken and Glowacki proposed a binary classification system for vascular anomalies based on natural history, cellular turnover, and histology [1,2,3].

The International Society adapted the basic binary system of classification of the vascular anomalies for the Study of Vascular Anomalies (ISSVA) [4,5,6,7]. The modified ISSVA system allows a systematic approach to vascular lesions that predictably correlates the clinical history, disease course, and treatment options, making the system clinically useful. The modified system incorporates the radiological studies like Doppler, MRI and angiograms, histopathological and histochemical studies in predicting the types and classification.

Table 1: Comparison between Malformations and Vasoproliferative lesions

Features	Malformations	Vasoproliferative lesions
First appearance	birth	After birth
Growth	Continue to adulthood	Limited growth
Static phase	No static phase	Static phase present
Involution	No involution	After growth it involutes
Stimulating factors	Trauma, surgery, oral pills, hormones	No effects

The International Society for the study of Vascular Anomalies (ISSVA) classification system divides vascular anomalies into two primary biological categories: (1) vasoproliferative or vascular neoplasms and (2) vascular malformations. The major distinction between the 2 categories is the presence of increased endothelial cell turnover in the former [6].

Congenital vascular malformations (CVM) are made of dysplastic vessels with no cellular proliferation. The vascular malformations will have structural abnormalities of the capillary, venous, lymphatic, and arterial channels that grow in proportion with growth of the child [5,6,7,8]. This is the main reason why the lesions should be excised early in childhood.

The next factor of importance is the flow rate. The high flow malformations are arteriovenous malformations and arteriovenous fistulas. The low

flow malformations are morphologically capillary and venous malformations. These have components of vascular, lymphatic and hamartomatous components. Those hamartomatous lesions may enlarge with pain and limitation of movements of the joints. The pure capillary and venous malformations will spread locally producing mass effect, local invasion and secondary changes like ulceration and bleeding. Low flow group have the property of platelet segregation. Hence we suggest surgical removal of the low flow malformation in early childhood.

Frequently many vascular centers treat low flow vascular malformations by sclerotherapy and embolisation. The procedure of sclerotherapy is done using materials like ethanol, sodium tetradecyl sulfate, and hypertonic saline. The results of sclerotherapy are unpredictable [9,10]. Prior to sclerotherapy, percutaneous phlebography is necessary to visualize vascular pattern inside the lesion and the flow into the adjacent vascular system [5]. The embolisation requires technologies like invasive vascular procedures (trans arterial or trans venous catheterization) especially in head and neck region, which is difficult in cases of children [13,14,15]. At times, sclerotherapy done for lesions of the limbs can produce thromboembolic complications. The thromboembolic necrosis of the lip and nose is not rare. The possibility of thromboembolic complications in head and neck regions cannot be neglected. Perioperative embolosclerotherapy has been performed in many centers, as preliminary preparation to reduce subsequent surgical morbidity. Yet another modality of treatment is steroid injections into the large lesions. The endothelial maturation process can be stimulated by the steroid, but the results are not satisfying. The intra lesion steroid therapy may help in reducing the problems of platelet segregations in large vascular anomalies (Kasabach Meritt phenomenon)[16].

Considering all, it is advisable to take decision on radical excision after studying the cases with Doppler, CT and/or MRI. The surgical management is performed in many ways. There are some reports on performing suturing of the vascular malformations all around. The aim of doing the suturing is to prevent bleeding and to reduce scarring and contractures [17]. In all other cases a multimodality and multidisciplinary approach is advisable [18]. If the lesions are localized and low flow anomaly, primary treatment option is total excision and primary reconstruction [19].

For performing the complete excision it is preferable to have magnification and finer dissection, to avoid complications like injury to small nerves or major vessels. The magnification can be a magnifying loop or an operating microscope. While dissecting the vascular malformation, the feeding or draining vessels can be localized with portable Doppler probe of 8MHz and 5 MHz These localized major feeders can be ligated first, lifting the mass towards its arterial feeder. The arterial feeders will be small and ligation will be easy. If the artery is primarily ligated the mass may reduce in size so that missing of some venous channels can occur.

Whenever dissection is done in areas like parotid the facial nerve should be identified first and then only proceed for excision of the malformation. The technical planning is done depending upon the site of lesion and main structures in that region. Referring to the other reported literature, the radical excision of the low flow malformations will give a cure with fine aesthetic result.

Conclusion

Early resection of low flow vascular malformations give better cure rate with good cosmetic correction and functional achievement.

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