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Therapeutic Strategies In Visceral Angiodysplasias

D.A. Loose, J.R. Loose, J.H. Weber and P. Bihari

Center for Vascular Medicine and Center for Vascular Disturbances and Vascular Defects, Hamburg, Germany

Summary

Vascular malformations can be categorized from the point of view of anatomic regions: central, visceral, peripheral and associated. In each of them two characteristics can be distinguished: localisation and morphology. The localisations are: 1. extracranial located vessels, 2. thoracic organs, 3. abdominal organs and urogenital systems, 4. multiple visceral localisations. The morphology is predominantly arterial, venous, lymphatic, av-shunting and combined.

6% of all patients with vascular malformations showed up with visceral malformations. Out of 64 cases 32 had treatment by surgery, 10 by interventional radiological treatment and 22 cases were not yet treated. The long follow-up results of surgical and non-surgical treatment of 42 cases were good or excellent.

Introduction

The publications of Arruda (1973) and Belov (1978) propagate to look at congential vascular malformations from the point of view of anatomic regions. So four main types are differentiated (in the distribution of the frequency):

1. peripheral vascular malformations
2. central vascular malformations
3. visceral vascular malformations
4. associated vascular malformations

In each of these types two characteristics can be distinguished: Localisation and morphology.

As for visceral vascular malformations the localisations are:

A) Malformations of the brain vessels and extra cranial located vessels nourishing the brain
B) Vascular malformations of the thoracic organs and the urogenital system
C) Vascular malformations of the abdominal organs and the urogenital system
D) Multiple localisations of visceral malformations.

Adverting the Hamburg Classification (Belov at al. 1985; 1989) the morphology is

predominantly arterial
predominantly venous
predominantly lymphatic
predominantly av-shunting
predominantly combined.

The basis for a reasonably modern treatment is: excellent clinical, functional and angiographic diagnostics. The choise of surgical or non-surgical treatment depends on the morphology and the special localisation of the malformation. (Loose 1997, 2001; Loose et al. 1994, 2002). Two goals have to be taken care of:

1. to treat the vascular malformation and to improve the hemodynamic disturbances
2. to preserve the specific organ, linked with the malformed vessels.

Materials and Methods

From October 1, 1970 until September 20, 2002 a retrospective analysis of all patients with visceral malformations was performed with a special interest to the long follow-up results.

Vascular malformations of the brain vessels and extra-cranial located vessels nourishing the brain are not so rare as supposed. As they are not always symptomatic in childhood they are quite often detected in young adults only. As for example in a case of a kinking and coiling of the internal carotid artery (Fig. 1a). In this predominantly arterial malformation the treatment of choice is the segmental resection of the internal carotid artery at the bifurcation and the new insertion of the vessel (Fig. 1b) in order to reconstruct the vascular continuity.

Vascular malformations of the thoracic organs and the thoracic wall are related to the lungs or the heart in most instances. So most malformations are pertaining to heart or lung surgery. In addition sometimes the malformation pertains the mediastinal region or the thoracic wall. As e.g. in a 13-year-old boy at his right thoracic and axilliary

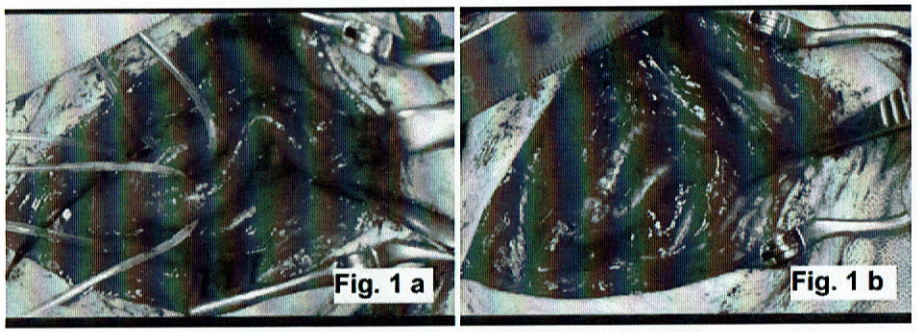

*Figure **1a**: Kinking and coiling of the internal carotid artery. **1b**: Situation after segmental resection of the internal carotid artery and new insertion of the vessel.*

region (Fig. 2 a and b) (opacification of the extent of the lesion by direct injection of contrast media)). The treatment of choice was the cutanectomy and total exstirpation of the venous malformation (Fig. 2c).

Vascular malformations of the abdominal organs and urogenital system occured e.g. in a 33-year-old woman. She complained of grow-

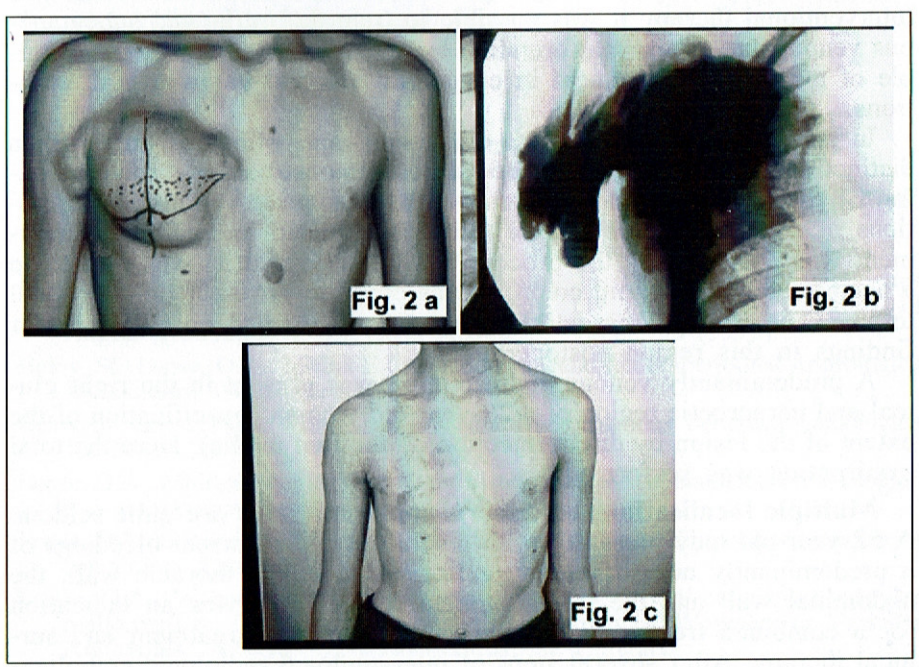

*Figure **2a**: 13-year-old boy with predominantly venous malformation of his thoracic wall. **2b**: Radiologic opacification of the extent of the lesion by direct puncture and injection of the media. **2c**: Aspect after surgery.*

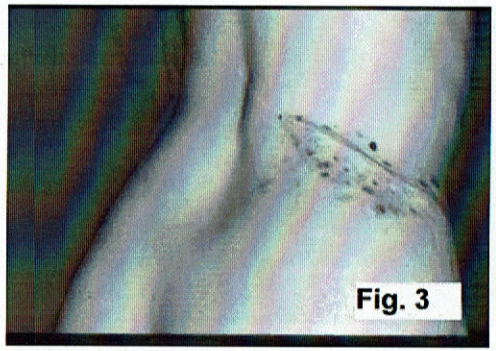

Figure 3: 33-year-old woman with intracutaneous blue spots in her right flank.

ing intracutaneous blue spots in her right flank (Fig. 3) combined with the feeling of undefined pressure in her right abdomen. The diagnostic examination demonstrated a predominantly venous malformation. There was a communication between the cutaneous findings and the center of the malformation, which was the right ovarian vein. The right renal vein and artery showed no pathology. In several interventional procedures the malformed veins of the right peritoneum were embolized. After this interventional therapy it was possible to treat the intra- and subcutaneous venous part of the malformation by surgery. This is a typical example of a combined treatment in congenital visceral vascular malformations.

In another case of a 20-year-old young man who presented with a distinct anemia the diagnostic procedures demonstrated a chronic bleeding of the sigmoid colon wall caused by numerous venous malformations in and around this part of the great gut. No interventional treatment was possible. As the process of the malformation was limited to a segment of the sigmoid colon, it was possible to resect this portional totally. The controll mesentericography demonstrated normal vascular findings in this region postoperatively.

A predominantly venous malformation was present in the right gluteal and paraproctic region of a 22-year-old woman (opacification of the extent of the lesion by direct injection of contrast media). Here the total exstirpation was performed.

Multiple localisations of visceral malformations are quite seldom. A 52-year-old lady presented with ulceration and recurrent bleedings of a predominantly arterio venous malformation of the thoracic wall, the abdominal wall and the left peritoneal region. This was an indication for a combined treatment: interventional radiologic treatment and surgical therapy. After several steps of interventional radiologic embolization treatment a stepwise surgical therapy was performed. By this stategy the hemodynamic disturbances could be diminished dramatically.

Results

In 32 years we registered that 6% of all patients with vascular malformations showed up with visceral malformations. A retrospective analysis was performed of 64 cases concerning the long follow-up results.

22 patients were not yet treated or refrained from treatment.

10 patients had an interventional radiologic embolization treatment alone or in combination with surgery.

32 patients had surgical treatment.

The subjective and objective long follow-up results were excellent in those cases where a total exstirpation of the malformation was possible and good in the cases where subtotal exstirpation of the malformation had been performed. In 5 cases further stepwise treatment by surgery was indicated.

Conclusions

In congenital vascular malformations each anatomic region need special therapeutic approaches. So it is worthwile to look at the different localisations of visceral malformations in order to standardise the tactic and the technique of treatment. The therapeutic opportunities are:

1. interventional radiologic embolization or
2. combined treatment (interventional and surgery) or
3. only surgery.

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