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MAIN TOPIC

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Surgical treatment of hemangiomas and vascular malformations in functional areas

Abstract Hemangiomas and vascular malformations (VM) in functional areas can be treated by a variety of methods. Because of the natural involution of hemangiomas, a non-aggressive approach is recommended. Active therapy is necessary only in cases where a function is affected such as vision, respiration, hearing, and feeding. Psychological problems can be an indication for early excision, and psychological/cosmetic reasons in the presence of fibrofatty tissue residues when the hemangioma has been involuted for late excision. In contrast to hemangiomas, no involution is to be expected for VMs, so that therapy depends mainly on the occurrence of functional problems and/or serious complications. Surgical excision still has a place, however, the indications are limited.

Key words Hemangioma · Vascular malformation · Surgical treatment

Introduction

According to Mulliken, vascular lesions can be classified as hemangiomas or vascular malformations (VM) based on their clinical and histologic characteristics [8, 11]. Hemangiomas are cellular tumors, exhibiting endothelial-cell proliferation. They are common occurring in about 12% of all children under 1 year of age. They typically and exclusively appear in early infancy, often beginning as a small macula and growing rapidly in the 1st to 4th weeks, then undergoing fatty replacement and involution by adolescence.

About 50% of all hemangiomas are involuted by the age of 5 years, about 100% by the age of 12. The histologic appearance depends on the stage of evolution: early lesions are characterized by proliferating endothelial cells and contain large numbers of mast cells. VMs are less frequently seen, in about 0.5% of all newborns. These lesions result from abnormal blood or lymphatic vessel morphogenesis. Histologically, VMs are characterized by normal endothelial cells and normal numbers of mast cells throughout their history. They are present at birth, but may not become clinically apparent until late infancy or childhood. Under the influence of hormones, trauma, and sepsis, changes may occur. Their growth is commensurate with that of the patient, and they do not involute. VMs are classified by the predominant type of vessel involved and include capillary (CM), venous (VeM), lymphatic (LM), and arteriovenous malformations (AVM). Another way of classifying vascular lesions is by their flow characteristics: hemangiomas, as well as CMs, VeMs, and LMs, are low-flow lesions while AVMs are high-flow lesions.

Differentiation between a hemangioma and a VM is very important for management and therapy, but may sometimes be quite difficult. The history and physical examination remain the most important diagnostic parameters. Ultrasound can be helpful; a duplex scan may aid in differentiating between low- and high-flow malformations. Magnetic resonance imaging (MRI) can also help differentiate between VeMs and hemangiomas [1]. The diagnosis of a VeM may be suggested by the presence of enlarged venous lakes and phleboliths. Arteriography remains the standard investigation for AVMs, but is useless for VeMs. Recently, experimental work has revealed that urinary levels of basic fibroblast growth factor, an angiogenic factor, are abnormally elevated during the proliferating phase of hemangiomas but return to normal during involution of the lesions or after therapeutic intervention. This may be a promising new method for distinguishing hemangiomas from VMs [4, 18].

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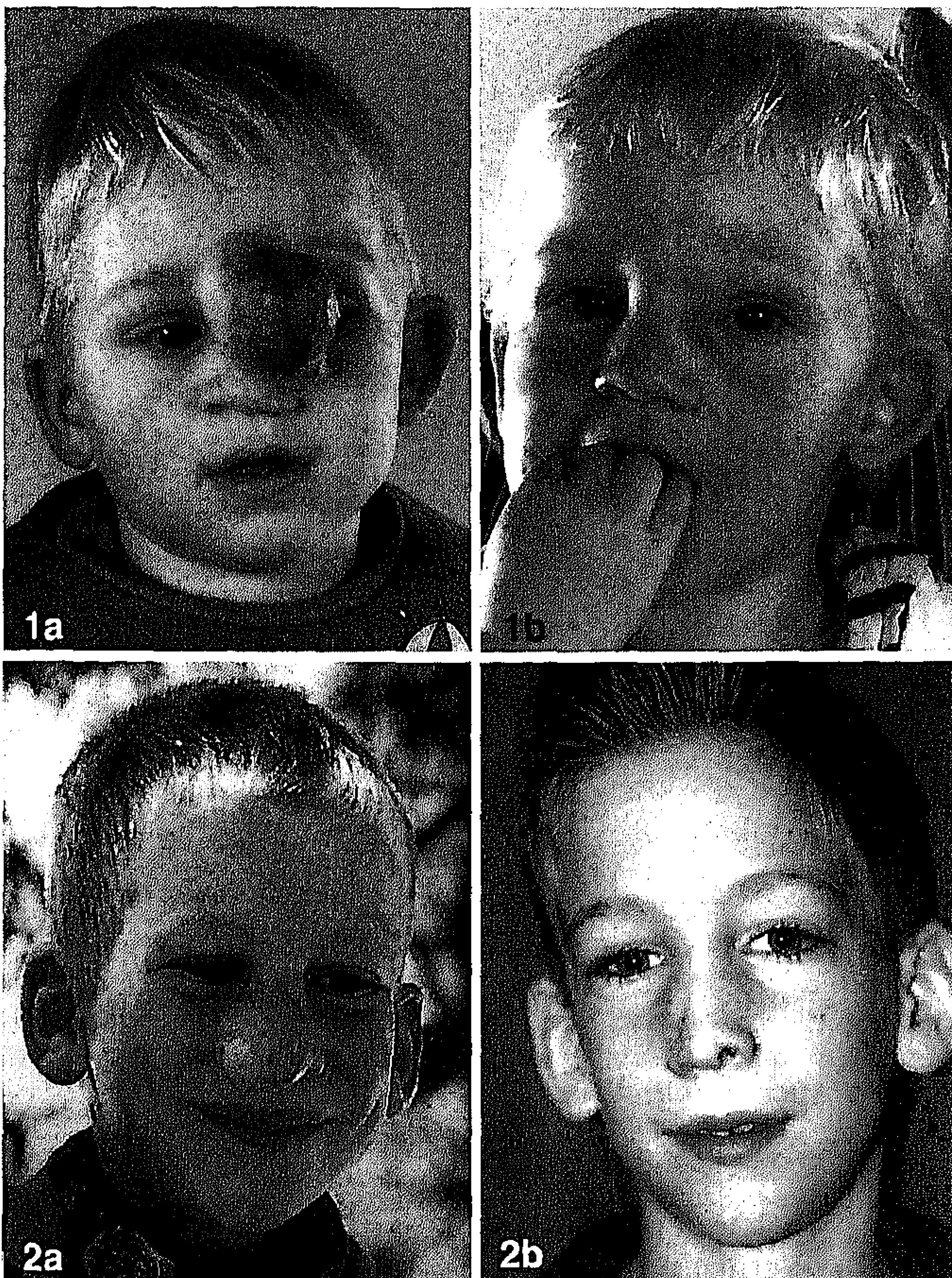


Fig. 1 a Fourteen-month-old boy with hemangioma on dorsal surface of nose. **b** After surgical debulking of hemangioma

Fig. 2 a Six-year-old boy with hemangioma of tip of nose. **b** After surgical debulking of hemangioma

Therapy of hemangiomas

In 1938, Lister was the first to publish a prospective study in which he stated that all fast-growing vascular tumors showed natural involution, on average at the age of 5 years [9]. Because of this natural involution of hemangiomas, a non-aggressive approach is recommended. In earlier years a variety of therapies were used: artificial ulceration, electrolysis, thermocauterization, radiotherapy, and compression. It is always important to tell the parents that surgery is not possible without scarring, while natural involution is generally followed by an inconspicuous scar. Treatment of

these benign lesions with radiotherapy is obsolete in view of the probability of developing skin cancer.

Active therapy is necessary only for complications or hemangiomas in special functional locations (eyelid, throat, mouth, etc.); growing hemangiomas at these locations sometimes interfere with vision, respiration, feeding, and hearing. Active therapy consists initially of corticosteroids, which can be given locally or systemically [14]; the success rate is reported to be in the range of 30% to 90% [11]. If corticosteroid therapy has failed, interferon can be used as a second-line treatment [3]. The penetrating ability of lasers limits their use in the treatment of hemangiomas [6, 17, 19]. Psychological problems in cases of severe disfigurement may be an indication for early surgical excision [10]. Cosmetically disturbing residual fibrofatty tissue and excess skin after involution of the hemangioma may be another indication for late surgical excision.

Case reports

Case 1 A 14-month-old boy presented with a hemangioma of the dorsal surface of the nose. No impairment of vision was noted. After carefully weighing the pros and cons, surgical debulking was performed mainly for psychological/cosmetic reasons. Primary closure was obtained (Fig. 1 a and b).

Case 2 A 6-year-old boy had a partially involuted hemangioma of the tip of the nose. No functional problems existed, but for psychological reasons the hemangioma was debulked and primary closure was carried out (Fig. 2 a and b).

Case 3 For psychological reasons, a hemangioma of the upper lip of a 5-year-old boy was treated by excision after ligation of the feeding superior labial artery (Fig. 3 a and b).

Case 4 A 14-month-old girl presented with multiple hemangiomas of the neck, shoulder, back, and buttocks. The hemangioma in the neck, which was small-based and protruding, was very distressing and therefore was simply excised. All the others were left alone for spontaneous regression.

Therapy of VMs

In contrast to hemangiomas, no involution is to be expected, so that therapy is mainly dependent on functional problems and/or serious complications: persisting pain, ulceration, and/or bleeding.

Therapy is differentiated according to the kind of malformation: CMs are best treated by laser therapy [6], while

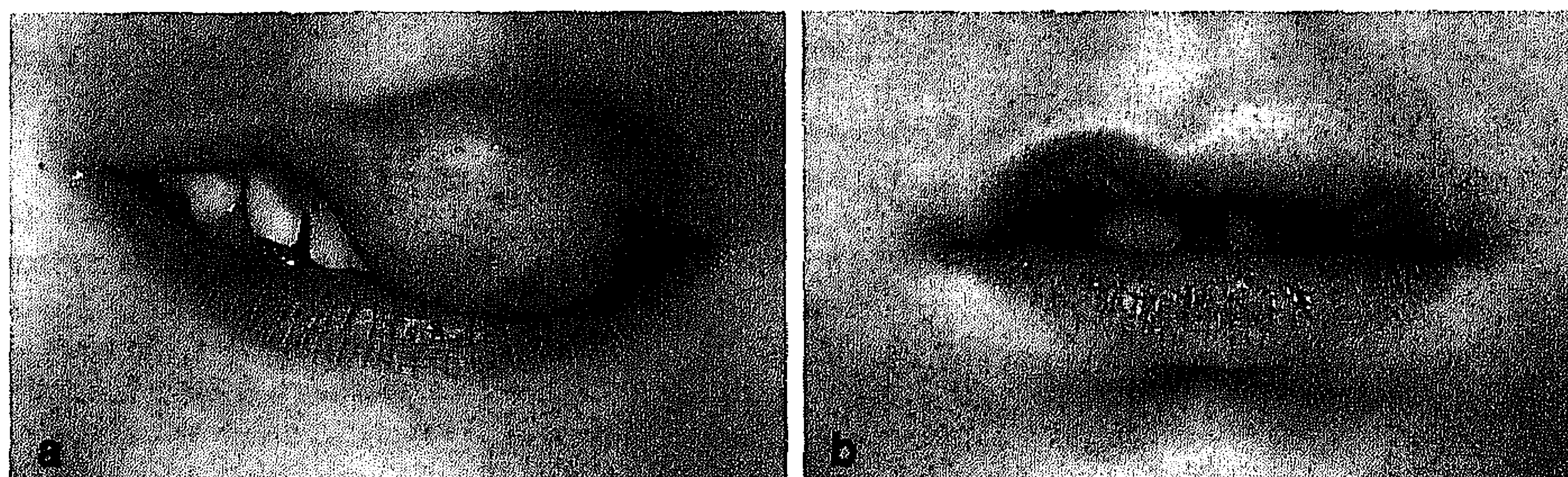


Fig. 3 a Five-year-old boy with hemangioma of upper lip. **b** After excision of lesion following ligation of feeding labial artery

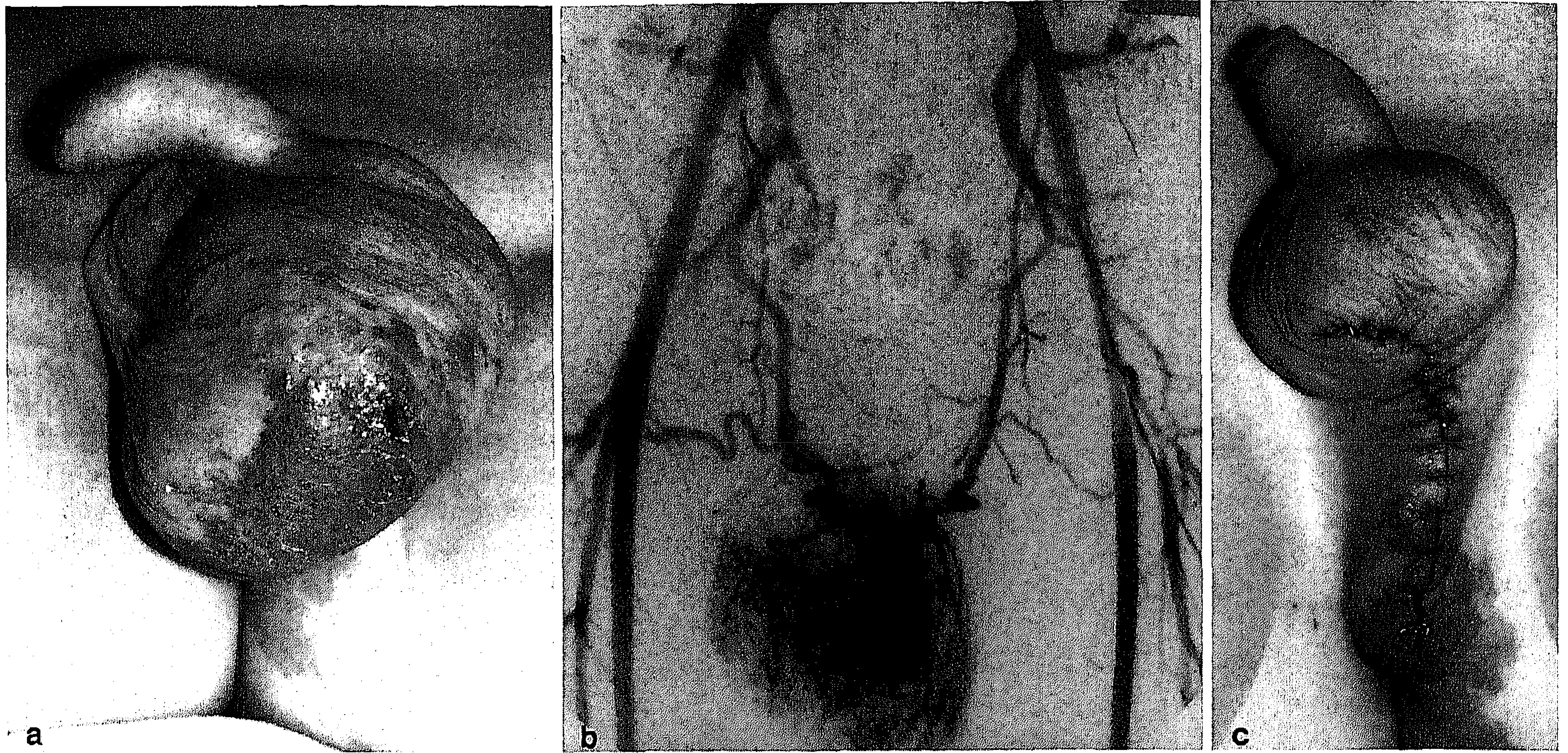


Fig. 4a Eleven-year-old boy with arteriovenous malformation (AVM) of scrotum. **b** Digital subtraction arteriography confirming diagnosis of AVM. **c** After embolization and excision of AVM

the main therapy for VeMs localized in the extremities consists of compression. Bulk reduction by means of sclerotherapy (alcohol, Ethibloc) or laser photocoagulation is often utilized in the head and neck region [4, 5, 15, 20]. Smaller, localized VeMs may better be excised. In our departments, cystic LMs are preferably treated surgically, although sclerosing therapies (Ethibloc, OK432) are also recommended by other authors [2, 12]. AVMs (high-flow lesions) are treated by embolization combined with surgical excision [16]. Surgical therapy alone leads to a high frequency of recurrence by means of collateral vessels. Ligation of the main vascular pedicle prohibits embolization in case of recurrence, so this is not recommended. Combination therapy is essential [7, 11, 13]; in large resections it may be necessary to reconstruct the defect that has been created with a free, revascularized tissue transfer [11, 20, 21].

Case reports

Case 5 An 11-year-old girl was referred because of congenital venectasias on the medial side of the lower leg. A small blue spot was observed directly after birth, which grew proportionally with the child. She later complained of a swelling that became painful 1 year before referral. On examination, a small mass with 2 cm in diameter was seen under the left knee. Medially, distal and lateral to this swelling, pretibial venectasias were observed. However, no signs of venous insufficiency or edema were discernible. The swelling had a normal temperature and no thrill was noted. A duplex scan suggested an AVM, while the clinical picture suggested only a local VeM. This was confirmed by additional MRI/MR angiography (MRA). Because of her complaints, the decision was made to excise the abnormal varix and perform sclerotherapy for the remaining venectasias. Histologic examination revealed a VeM with one recent and multiple older thrombi. After the operation the patient was asymptomatic.

Case 6 An 11-year-old boy presented with a large swelling of the scrotum, which had existed since birth but had grown rapidly during the last 12 months. Ulceration and bleeding occurred regularly.

Inspection revealed a large capillary malformation of the scrotal and perineal region. In addition, a large swelling was seen in the scrotum. Duplex and digital subtraction arteriography confirmed the diagnosis of an AVM. Embolization and resection was carried out; because of the proximity of the anal sphincter, the resection in the perineal region was incomplete. The postoperative recovery was uneventful. At 4-year follow-up increased bulging of the perineal area was seen. Although the scrotal area remains free, the perineal skin again shows ulceration. MRI/MRA and angiography was again performed and showed a recurrence of the AVM in the perineum. Because of increasing physical and psychological problems, it was decided to embolize and resect in the near future (Fig. 4a-c).

In conclusion, it should be borne in mind that only a very small percentage of patients with hemangiomas require active intervention, however, in a small subpopulation surgical excision may be an appropriate and justified treatment. These cases include endangerment of vision, hearing, respiration, or frequent bleeding, as well as psychological problems because of severe disfigurement of the face. VMs occur less frequently, and in many instances no treatment or conservative management is preferred. Among several therapeutic modalities, surgery still has a place in localized VeMs and cystic LMs and in combination with embolization for AVMs.

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