GIANT HEMANGIOMA OF THE THIGH
- CASE REPORT

SUMMARY:
Hemangiomas are the most common vascular tumor of infancy. Affecting approximately 4% of children they are benign endothelial cell neoplasm. Usually, they are not fully present at birth, appear in infancy and have a natural history of proliferation and involution. We report the case of a male infant who had a large hemangioma involving two-thirds of the right thigh. MRI with 3D reconstruction revealed a 10/7 cm tumor localized at the inguinal area and the proximal 2/3rd of the right thigh between the sartorius and rectus femoris muscle. Surgery intervention was undertaken and total resection of the tumor was performed.

Key-words: hemangiomas, thigh, vascular malformations, tumor, sclerotherapy, surgical excision

HEMANGIOM GIGANT DE COAPSĂ LA COPIL - PREZENTARE DE CAZ

REZUMAT:
Hemangioamele sunt cele mai frecvente tumori vasculare ale copilului. Afectează aproximativ 4% din copii, fiind tumori benigne ale celulelor endoteliale. De obicei, ele nu sunt prezente la naștere, apar în copilărie și au o istorie naturală a proliferării și involuției. Prezentăm cazul unui copil de sex masculin, care are un hemangiom mare care cuprinde două treimi din coapsa dreaptă. RMN cu reconstrucție 3D a relevat o tumoră de 10/7 cm localizată în zona inghinală și 2/3 proximale ale coapsei dreapte între mușchiul sartorius și mușchiul drept femural. După intervenții chirurgicale repetate s-a reușit excizia totală a formațiunii tumorale.

Cuvinte cheie: hemangioame, malfomații vasculare, scleroterapie.

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BACKGROUND

Vascular anomalies are a heterogeneous group of entities having a large clinical spectrum of lesions ranging from small skin stains to large life-threatening lesions. Treated inconstantly by various medical specialties, there is a general confusion regarding nomenclature and classification. First comprehensive classification was made by Mulliken and Glowacki based on distinctive clinical, radiologic, and histological findings (1). It categorized vascular anomalies as either hemangiomas or malformations and later on, in 1996 it was adopted by the International Society for the Study of Vascular Anomalies. Hemangiomas are the most common vascular tumor of infancy. Affecting approximately 4% of children they are benign endothelial cell neoplasm. Usually, they are not fully present at birth, appear in infancy and have a natural history of proliferation and involution (2).

Unlike hemangiomas vascular malformations are structural anomalies that occur as a result of errors in embryogenesis. In contrast to the proliferative nature of hemangioma endothelium, the endothelium of vascular malformations has normal cellular turnover (3). They are histologically normal, but with abnormal architecture derived from arteries, capillaries, veins, lymphatics or a

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combination of them. Most often, the lesions are present at the time of birth and grow proportionately with the child (4). They occur in any locations and are in most cases single (4). Venous malformations are the most common. Complications include: esthetic prejudice, local compression and displacement of adjacent organs, haemorrhage and ulceration. General complications are: cardiac failure secondary to arterio-venous shunting, coagulopathy due to platelet trapping (Kasabach- Merritt syndrome) and/or consumption of clotting factors (disseminated intravascular coagulation) and infection (5). Treatment options include surgery but complete surgical excision is often not possible. Sclerotherapy alone or combined sclerotherapy- surgical excision is performed when complete surgical excision is not possible.

**CASE REPORT**

A male infant was born through cesarean section at 39 weeks gestation after an uncomplicated pregnancy. Birth weight was 3200g. Apgar score was 8 at 1 and 9 at 5 minutes. He was the first child of nonconsanguineous parents and had no significant medical history in family. Clinical exam at birth revealed a large hemangioma involving two-thirds of the right thigh (fig.1).

After the first month of life focal necroses arise and intermittent bleeding starts from the center of the tumor (fig.2).

He was admitted in our hospital at the age of 6 weeks. Extended laboratory tests were performed: HGB 8.7g/dl, RBC 3,2x 106, PLT 442x 103/ mm3, WBC 21.2 x103/mm3, VSH of 50 and PCR: 29.17 mg/l. The lab tests for hepatic and renal function were in normal range. Chest X-ray was normal. MRI with 3D reconstruction revealed a 10/ 7 cm tumor localized at the inguinal area and the proximal 2/3rd of the right thigh between the sartorius and rectus femoris muscle. The arterial sequence was normal. The tumor filled with contrast substance during the venous phase and showed numerous vascular spaces communicating with the femoral vein. The major communication was through a 5 mm thick vessel located near the inguinal ring on the intern aspect of the femoral vein (fig.3).

The right iliac vein was dilated. The inferior cava, left iliac and femoral vein were normal. Surgery intervention was undertaken and subtotal resection of the tumor was performed. Because the lesion occupied most of the right thigh it was decided that the resection should be made in more stages in order to avoid wound closing issues. Even so wound closure was made with great difficulty putting considerable tension in the margins (fig. 4).

**Fig. 4**

After surgery antibiotics, antinflammatory drugs and pain killers were administered. Five days after the operation the surgical wound becomes dehiscent (fig.5).
Local treatment with antiseptics and daily wound dressing were applied. After another 14 days the surgical wound was closed and only a small part of the tumor was left behind for step two (fig.6).

The patient was discharged with most of the tumor removed. A relatively large portion was left on the interior aspect of the thigh. On the anterior aspect remained only a thin marginal portion (fig. 7).

The patient was readmitted in our hospital after two months and a second surgical intervention was performed. A remained 3/2 cm tumoral portion from the interior aspect of the thigh was removed (fig. 8).

Postoperative course was uneventful and the patient was discharged 5 days from surgery.

The remained portions and part of the scar were surgically removed in two steps at 2 months interval (fig.9).

At the age of 1 year, the patient was free of tumor. The remaining scar is set for surgical excision 12 months after the last surgical intervention (fig. 10). Six months later the patient was fully recovered, the scar diminished in size and had no walking impairment (Fig.11)
DISCUSSION

Management of large vascular tumors in children is difficult. It is important to identify the type of the lesions in order to decide the best treatment options. Differentiation between hemangiomas and vascular malformation is essential because the treatment is fundamentally different. For hemangiomas, even the large ones, intervention should not be attempted unless complications occur (4). Most of them involute after the first year of life (4). Vascular malformation does not involute and sooner or later a kind of intervention is required. Despite initially presumed to be a hemangioma, the main clinical and imagistic features indicated a venous vascular malformation as described by Burrows et al (6). This was later confirmed by the pathological exam. MRI was by far the most useful imaging tool. Using 3D reconstruction software we were able to see the lesions in detail and its rapport with the surrounding healthy tissues and main vessels. Identifying all large vessels that supply the tumor prior to surgery is a huge benefit in planning it, because blood loss in an already anemic infant is significantly lower.

Complete surgical excision is ideal for vascular malformations (3). The timing for treatment is determined mainly by the presence of the complications (4). Early surgical excision was decided because of the necrosis, infection and haemorrhage leading to severe anemia. Other treatment options like sclerotherapy, compressive bandage, ligation of the pedicle or corticosteroids were inadequate due to the complications. Unfortunately, the largeness of the lesion made it impossible for us to remove it all in one step.

Fig. 9. At the age of 12 months the patient was free of tumor.

Fig. 10. At the age of 1 year, the patient was free of tumor.

Fig. 11. The patient at the age of 1 year and 6 months. No walking impairment was present due to the remaining scar.
Our attempt in the first operation to remove a large part of the tumor was followed by deficient wound closure and infection. The lesion was located in a region with poor hygiene conditions that contributed to the postoperative wound infection. Complete surgical excision of the lesion was possible only after several steps. Because the tumor occupied more than 2/3 of the thigh we were unable to close the wound using pediculated flaps. Free flaps were inadequate because of such a large defect and because of the profound alteration of the local architecture of the vessels.

CONCLUSIONS

Identification of the type of vascular malformation is essential. The timing for treatment is determined mainly by the presence of the complication. Good preoperative images of the tumor are the most useful tools for careful planning and could represent the key for the success of the intervention.

References: