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CASE REPORT

Fibro-adipose vascular malformation (FAVA) in forearm: a successful surgical approach and literature review

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Abstract

Fibro-adipose vascular anomaly (FAVA) is a new entity characterized by fibro-adipose infiltration of muscle, pain, and contracture. The main differential diagnosis includes venous malformation and intramuscular hemangioma. Early treatment and physical therapy are of paramount importance to prevent severe contractures, muscle fibrosis, and limb atrophy. We present a case report of a 24-year-old female patient with FAVA in the volar aspect of left arm, presented with limitation to pronation and supination and pain. The patient was operated in 2017 in our center, performing a total resection of the mass and 2-year follow-up. We describe the criteria that we performed to diagnose the left forearm FAVA in this patient, based on the history; the magnetic resonance imaging, which demonstrated muscle replaced by heterogeneous soft tissue, intramuscular phlebolit, and dilated vessels; and histological images that showed a vascular proliferation associated with adipocytes and dense fibrous tissue replacing and infiltrating the skeletal muscle. En bloc resection of the mass was performed successfully, and it gave excellent results with recovery of limitation and improvement of pain and swelling that were maintained at 2-year follow-up. Surgery is a reliable and curative option in FAVA and could be the only one for long-term control of this entity.
Level of evidence: Level V, therapeutic; diagnostic study.

Keywords FAVA · Vascular anomaly · Surgical treatment · Forearm

Introduction

Fibro-adipose vascular anomaly (FAVA) is a new delineated disorder included in the International Society for the Study of Vascular Anomalies of 2018, as a provisionally unclassified vascular anomaly. It is associated with pain, contracture of affected extremity, and functional limitations [1], explained by fibro-fatty infiltration of muscle and unusual phlebectasia, constituting a distinct clinical, radiological, and histopathological entity. FAVA most commonly occurs on the calf of middle-aged women, and the main differential diagnosis includes venous malformation and intramuscular hemangioma [2].

Early treatment and physical therapy are of paramount importance to prevent severe contractures, muscle fibrosis, and limb atrophy. Optimal treatment for FAVA has not been fully established due to its recent description. Treatment options have included sclerotheraphy, cryoablation, surgery, or even medical treatment [3].

Case report

We present a case report of a 24-year-old female patient with a 10-year history of a progressive painful mass in

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volar aspect of the left forearm. The patient was clinically diagnosed of venous malformation without histopathology and treated with sclerotherapy at another center, but without control of pain and progression of the mass. Finally, she was referred to our center.

At examination, there was a palpable mass in the forearm with swelling, important limitation to pronation and supination, atrophy of the thenar, hypothenar, and interosseous muscles. Magnetic resonance imaging demonstrated a 12 × 4 × 4 cm mass in the volar aspect of the left forearm. In axial T1-weighted MRI, there was muscle replaced (flexor carpi radialis and flexor digitorum superficialis) by heterogeneous soft tissue with signal intensity higher than adjacent normal muscles. Coronal T1-weighted MRI noted intramuscular phlebolit. Sagittal fat-suppression MRI noted the longitudinal distribution of disease and dilated vessel (Fig. 1).

Due to the functional limitation and pain, surgery was proposed, and an en bloc resection was performed. Operative exploration evidenced a fibrotic mass extending into the adjacent fascia and fat planes, in contact with the radial and cubital bone but separated by a delimited plane. The muscular and bone component of the mass was resected completely, without neurolysis (Fig. 2).

Histological examination (Fig. 3) showed a vascular proliferation associated with adipocytes and dense fibrous tissue replacing and infiltrating the skeletal muscle. The vascular component included clusters of muscularized venous structures intermingled with ectatic thin-walled venous channels and capillary vessels organized in lobular pattern. No organized thrombi were observed. Some of the vascular lobules were surrounded by lymphoplasmacytic aggregates. Lymphatic vascular component was supported by endothelial D2-40 immunopositivity. Furthermore, focal bone metaplasia, atrophic skeletal muscle, and entrapped nerves were also noted.

Follow-up examination 3 months after surgery evidenced an important improvement in pronation and supination, with recovery of limitation and improve of pain in 2-year follow-up.

Discussion

Traditional classification for low-flow vascular malformations includes venous malformation (VM) and lymphatic malformation (LM) [4]. However, FAVA is a newly recognized clinicopathologic and radiologic entity, included in the International Society for the Study of Vascular Anomalies of 2018 as a provisionally unclassified vascular anomaly.

In 2014, Alomari et al. described a cohort of 18 patients with clinical, radiologic, and histopathological features of this new vascular entity [2]. This was

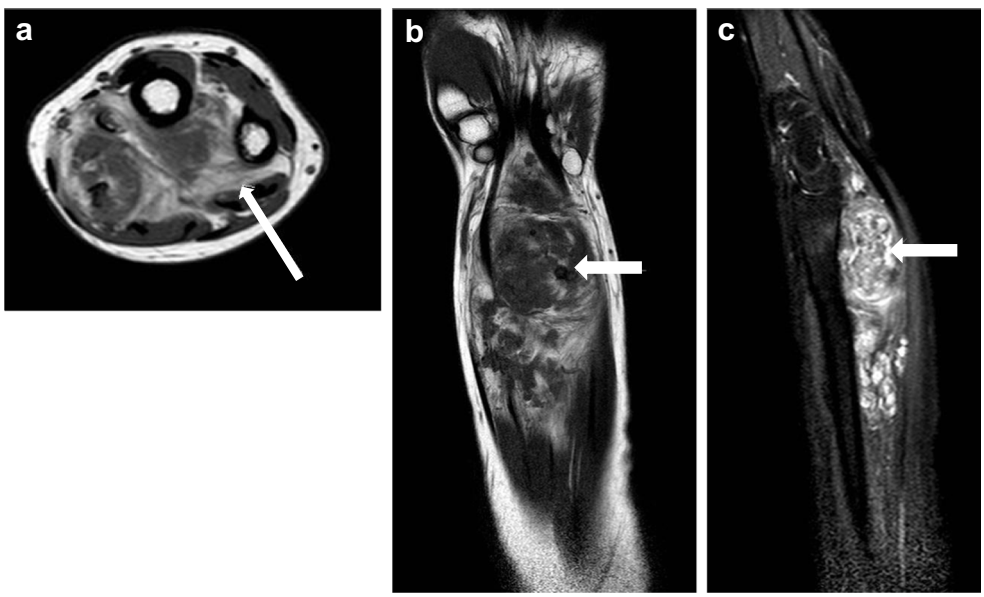

Fig. 1 Magnetic resonance imaging (MRI) of the left forearm evidenced a diffuse fibro-adipose vascular anomaly (FAVA) in the volar aspect. **a** Axial T1-weighted MRI. Muscle replaced by heterogeneous soft tissue with signal intensity higher than adjacent normal muscles (white arrow). **b** Coronal T1-weighted MRI, note intramuscular phlebolit (white arrow). **c** Sagittal fat-suppression MRI, note the longitudinal distribution of disease and dilated vessel (white arrow)

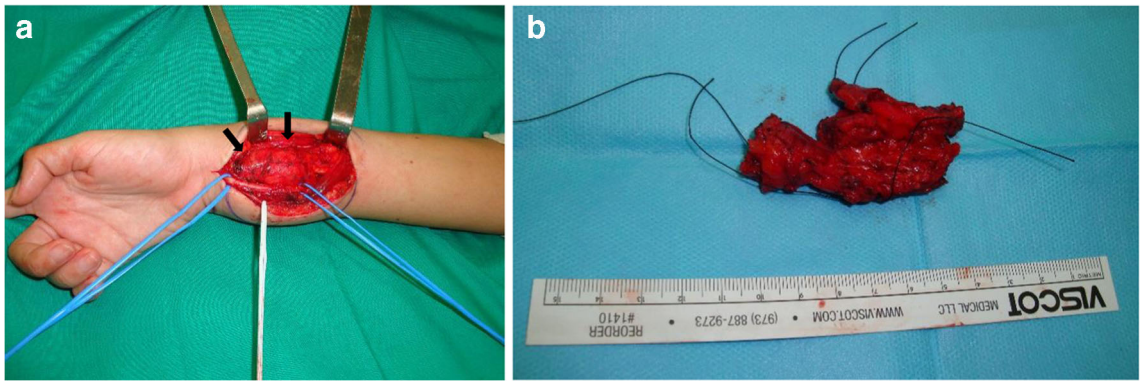

Fig. 2 Intraoperative findings. **a** Left forearm infiltrated in the flexor aspect (flexor carpi radialis and flexor digitorum superficialis) by fibro-adipose tissue (black arrow). **b** Specimen of fibro-fatty accumulation after resection

categorized like a complex mesenchymal malformation that includes fibro-fatty infiltration of the muscles, phlebectasia and intertwining of anomalous veins, and subcutaneous and cutaneous lymphatic malformation [1].

The clinical features of FAVA include unusual and disproportionately severe pain and contracture, affecting specific anatomic areas (calf and forearm). The constant pain and discomfort seen in these patients are multifactorial. There is a dysfunction and muscular contractility affectation explained by fibro-fatty infiltration of the muscles. The neural and perineural fibrous scarring contributes to the neurogenic pain. The anomalous ectatic veins have stasis and hypertension causing pain, swelling, and discomfort. Pain explains loss of movements of

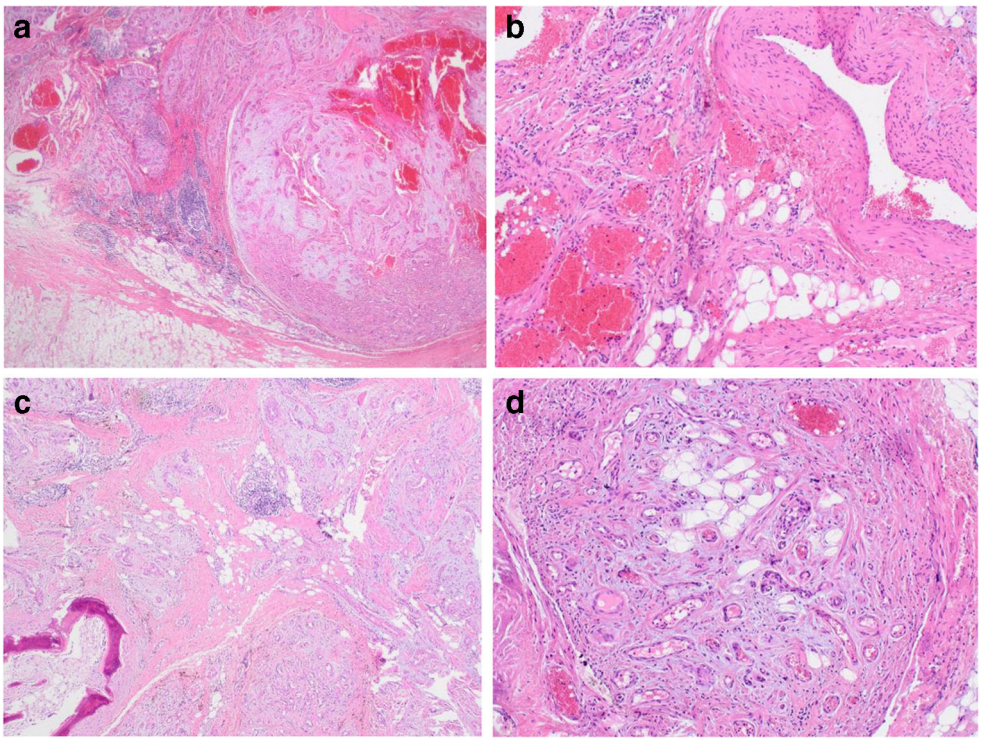

Fig. 3 Histopathologic examination. **a** Variable admixture of dense fibro-fatty tissue and vascular clusters, with ecstasies. Lobular pattern of growth and lymphoplasmacytic aggregates (H&E, × 20). **b** Clusters of thin-walled, blood-filled venous channels alternated with muscularized vessels in fibro-adipose tissue (H&E, × 100). **c** Lobular pattern of growth and lymphoplasmacytic aggregates. Focal bone metaplasia (H&E, × 40). **d** Thin-walled channels and adipose tissue embedded in a fibromyxoid stroma (H&E, × 100)

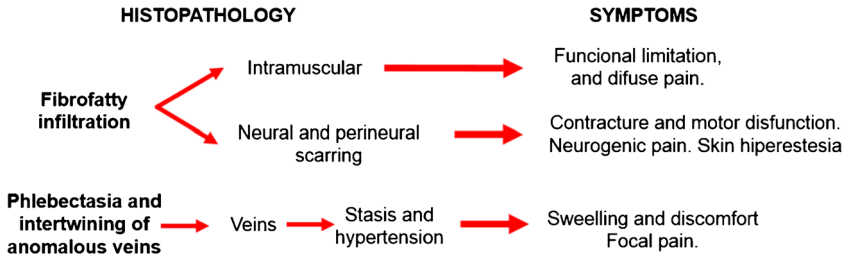

Fig. 4 Physiopathology of FAVA [1, 2]

involved extremity, contributing further to dysfunction, constituting a vicious cycle of pain and contracture [1] (Fig. 4).

Similar to the cohort reported by Alomari et al. in 2014, our patient was a young woman who presented with a painful forearm mass, that was initially diagnosed as a VM with muscle involvement on MRI/ultrasound, and firstly treated with sclerotherapy.

On MR imaging, FAVA is of heterogeneous signal intensity on T1-weighted images and of moderately hyperintense, heterogeneous signal intensity on T2-weighted images. It is centered in the musculature, frequently with associated increased fat in the fascial and subcutaneous regions and readily identifiable phlebectasia [5] replacing the normal muscle [2] and sometimes complicated by thrombi [6].

Among the histological findings, similar to other cases previously reported [2, 7], our case showed vascular proliferation associated with adipocytes and dense fibrous tissue replacing and infiltrating the skeletal muscle. The vascular component included clusters of muscularized venous structures intermingled with ectatic thin-walled venous channels and capillary vessels organized in lobular pattern. The presence of phleboliths is more likely to represent venous malformations; however, it is occasionally present in FAVA as in our case [2]. The main differential diagnosis included a venous malformation [2] (see Table 1).

Treatment options for this entity include observation/conservative approach, physical therapy, casts or splints, sclerotherapy, steroid injections, surgical excision, neurolysis or neurectomy [1], cryoablation, and more recently, the use of sirolimus [3].

Sclerotherapy could be performed in the smaller venous component, with less effect in dominant solid fibro-fatty component [2], which explains the poor management of this disease with this treatment.

Shaikh et al. in 2016 reported the percutaneous cryoablation in a retrospective cohort study of 20 patients and 26 sites, concluding that it is a safe and minimally invasive option for symptomatic FAVA lesions. Eighty percent of the patients were satisfied with this treatment, but control of

Table 1 Differential diagnosis between FAVA and venous malformation [1, 2]

Features	FAVA	Venous malformation
Location	Calf and forearm	Quadriceps > gastrocnemius
Symptoms	Disproportionately and severe pain. Focal, diffuse, or neurogenic. Contracture and functional limitation.	Episodic pain or asymptomatic.
Ultrasonography	Solid, hyperechoic muscle with dilated veins.	Hypoechoic, septated, and compressible spaces.
MRI	Solid, heterogeneous, diffuse or moderate no strong enhancement, phleboliths.	Typical fluid signal, no major solid component. Heterogeneous postcontrast enhancement.
Histopathology	Fibrosis, vessels fat, slow-flow vascular malformations (venous > lymphatic) within the muscles, and adjacent subcutaneous tissue. Venous channels with phlebectasia.	Vessels walls Venous channels spongiform, less phlebectasia.

FAVA, fibro-adipose vascular anomaly; *MRI*, magnetic resonance imaging

pain, swelling, and functional restriction was achieved in less than 50% of patients [1].

In 2019, Ramaswamy R et al. reported the use of cryoablation as a primary line treatment for 4 cases of FAVA in the foot and calf with control of pain in 3 patients. They concluded that percutaneous cryoablation appears to be safe and effective in the treatment of symptomatic low-flow vascular malformations, including FAVA, either as a primary modality or secondary therapy in addition. However, they only use this procedure in circumscribed focal lesion and localized in muscle and/or subcutaneous region [8].

In 2017, Erickson et al. presented two patients affected with FAVA with severe pain and functional impairment in which sirolimus produced rapid, dramatic improvement. They hypothesized that FAVA may be the result of a somatic activating mutation in PIK3CA, a gene related to overgrowth spectrum that includes a group of disorders with fibro-adipose overgrowth and infiltration into muscle and vascular malformations. They concluded that oral sirolimus was rapidly effective in those two patients, with improvement in the quality of life. On the other hand, they anticipate there will be recurrence after cessation of treatment and a very low dose maintenance therapy would be necessary [3] with the concomitant risk of immunosuppression.

Surgical resection is associated with longer recovery, increased risks, and sequelae [1], but is an effective treatment and the only one for long-term control of FAVA [2]. In cases with extensive muscle involvement and neurovascular entrapment, compartment decompression, partial resection, neurolysis, and tendon lengthening can be very effective [9]. Occasionally, the entire muscles or muscle groups may need to be removed to treat the pain and contracture; in these cases, tendon transfers or free functional muscle reconstruction is often necessary [10]. In our case, surgical resection gave excellent results with recovery of limitation and improvement of pain and swelling that were maintained at 2-year follow-up. There are few cases in literature about surgical treatment of forearm FAVA and therefore, additional studies in a larger cohort are needed to evaluate different management. [1, 2, 7, 9].

Conclusion

To our knowledge, this article presents a case of successful surgical treatment of FAVA in forearm, without recurrence at 2-year follow-up and without sequelae. We consider that surgery is a reliable and curative option in selected cases and could be the only one in for long-term control of FAVA; although, larger studies are needed comparing different treatment options for this disease.

Author's contributions All authors contributed to the study conception and design. Material preparation, data collection, and analysis were performed by Susana López, Andreé Ibarra, Eulalia Baselga, Carmen Vega, Caterina Fumagalli, Sandra Valverde, and Jaume Masia. The first draft of the manuscript was written by Andreé Ibarra, and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Statement of informed consent Informed consent was obtained from the individual participant included in the study. Informed written consent was obtained from the patient in this case report for inclusion of clinical data and photographs.

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