

Treatment of Acquired Digital Arteriovenous Malformation with Progression during Pregnancy

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Summary: Acquired digital arteriovenous malformation (AVM) is a relatively rare form of AVM that occurs in the fingers. Together with a review of the literature, we report a rare case of acquired digital AVM that enlarged after pregnancy. The patient was a 32-year-old woman with the chief complaint of digital swelling accompanied by a burning sensation. During her pregnancy, the swelling of the fingers recurred, with a symptom of throbbing sensation. After giving birth, the swelling reduced but did not completely disappear. At the first visit, we observed purple discoloration and swelling of the ulnar aspect of the proximal interphalangeal joints of the left hand. Contrast-enhanced computed tomography scanning via the digital arteries of the left hand revealed a lesion showing early venous return, leading to the diagnosis of AVM. Surgery was performed under general anesthesia. The digital artery supplying the lesion was identified and dissected under a surgical microscope. At 15 months after surgery, there was no recurrence, sensory dysfunction, or mobility impairment. (*Plast Reconstr Surg Glob Open* 2019;7:e2535; doi: 10.1097/GOX.0000000000002535; Published online 12 December 2019.)

INTRODUCTION

Acquired digital arteriovenous malformation (AVM) is a relatively rare form of AVM that occurs in the fingers.¹⁻⁴ Here, we report a rare case of acquired digital AVM that enlarged after pregnancy together with a review of the literature.

CASE REPORT

This study was approved by the appropriate ethics committee and has therefore been performed in accordance with the Declaration of Helsinki (1964) and subsequent amendments.

The patient was a 32-year-old woman with the chief complaint of digital swelling accompanied by a burning sensation. There was no remarkable medical history or family history. Linear erythema was first noted on the left fingertip when the patient was 13 years old. During her first pregnancy at the age of 28 years, the patient became aware of a burning sensation and swelling of the fingers, which gradually progressed. After giving birth, the swelling

resolved and she was followed up for observation. During her second pregnancy at the age of 30 years, the swelling of the fingers recurred, with a new symptom of throbbing sensation. After giving birth, the swelling reduced but did not completely disappear, and so she was referred to our clinic for further examination.

At the first visit, we observed purple discoloration and swelling of the ulnar aspect of the proximal interphalangeal joints of the left hand. There was no sensory dysfunction of the fingertips or impaired mobility of the fingers (Fig. 1A). Contrast-enhanced computed tomography scan via the digital arteries of the left hand revealed a lesion showing early venous return, leading to the diagnosis of AVM (Fig. 2). Surgery was performed under general anesthesia and tourniquet application. The ulnar digital neurovascular bundle was separated under a surgical microscope. The digital artery supplying the lesion was identified and dissected after ligation; the digital nerves were preserved (Fig. 1B). Pathological examination revealed relative thickening of the blood vessel walls, and numerous irregular vessels were observed, including those with branching appearance (Fig. 3). As of 15 months after surgery, there was no recurrence, sensory dysfunction, or mobility impairment. Complete resection of the left ulnar lesion was confirmed by postoperative contrast-enhanced computed tomography scan.

DISCUSSION

The term “acquired digital AVM” was first used by McCulley et al. to describe a condition characterized by a pulsatile mass associated with purpura in the fingers

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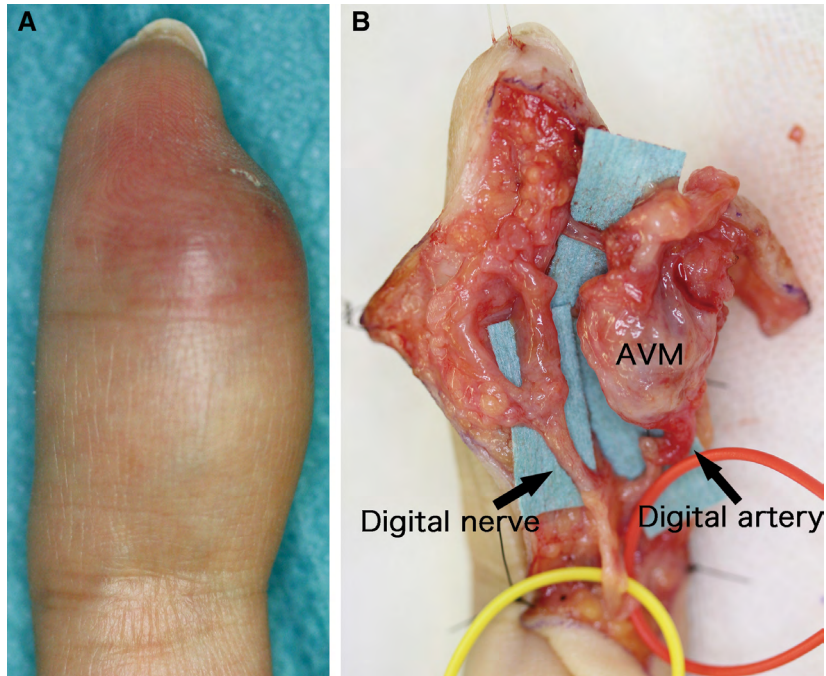


Fig. 1. Clinical findings at first visit and intraoperative findings. A, Initial examination confirmed purple discoloration and swelling of the ulnar aspect of the distal side of the left proximal interphalangeal joint. Fingertip sensory function was normal, and there was no mobility impairment. B, Surgery was performed under general anesthesia with tourniquet application to separate the ulnar digital neurovascular bundle. The digital artery supplying the lesion was identified, and the lesion was dissected after ligation; the digital nerves were preserved.



Fig. 2. Preoperative computed tomography scans processed with maximum intensity projection.

in pregnant women.⁵ To our knowledge, only 45 cases of acquired digital AVM have been reported to date. The incidence rate has not yet been reported in any studies. Treatment using primary suture, skin grafting, and laser irradiation has been reported with favorable outcomes; studies on recurrence have not yet been reported.⁵

AVM is an arteriovenous anastomosis that bypasses the capillary system, where arteries and veins are connected at the nidus. In the present case, the lesion enlarged in 2 consecutive pregnancies. During the second pregnancy, the disease progressed from Schöbinger Stage 1 to Stage 2. Given the possibility of further enlargement in a subsequent pregnancy, the patient was considered eligible for surgical treatment.

General mechanisms for AVM enlargement upon pregnancy are considered to be a combination of the following factors:

1. Increased estrogen and progesterone inducing vasodilation and angiogenesis that in turn increase blood flow and blood pressure.
2. Uterine contractions during labor leading to increased blood pressure.
3. Uterine enlargement leading to increased venous pressure.

It has been reported that the interplay of these factors increases shunt volume, leading to enlargement of the AVM. In our case in particular, it is thought that vascular changes were mainly attributable to hormonal factors.⁵⁻⁷

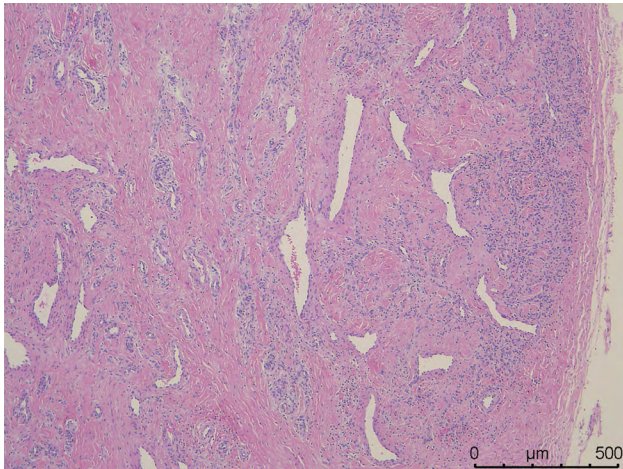


Fig. 3. Pathological examination showing proliferation of vessels with relative thickening of the vessel wall and irregular shapes with branching.

Kulungowski et al reported that increased expression levels of growth hormone receptor (GHR) occur in vascular malformation, especially in AVM, and that GHR is likely to be involved in enlargement of the lesions.⁸

In contrast, pituitary-derived growth hormone (GH) levels do not increase during pregnancy,⁹ although levels of human placental lactogen (hPL) secreted by the placenta are known to increase. hPL has GH-like effects but does not have a specific receptor and is reported to bind to GHR. A possible mechanism of AVM enlargement may be hPL binding to GHR that is highly expressed within the lesion.¹⁰

CONCLUSIONS

We encountered a relatively rare case of digital AVM that presented with enlargement during pregnancy. In patients with AVM who have a possibility of future pregnancy, the likelihood of disease progression during pregnancy must be considered, as well as the fact that treatment

options may be limited. Treatment strategies and timing must be adequately considered for these patients.

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