

Case Report

A Surgical Case of Venous Aneurysm of the Cephalic Vein with Unique Clinicopathological Findings for Venous Dissection

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Abstract

We presented an extremely rare case of a 38-year-old female's venous aneurysm of left cephalic vein with unique histopathological features, displaying variably thinned medial wall with focal, markedly reduced or absent smooth muscle cells and elastic fibers, most likely leading to the venous dissection with an intimal tear and many medial blood-filled vascular channels. We propose that those venous dissection-like findings would be a new feature especially from the clinicopathological viewpoints and might be considered in the classification of venous aneurysm. Further collecting cases are needed to validate the presence and significance of venous dissecting aneurysm as a new histopathological entity. This short report could interest the scientific community, taken together with potentially specific findings of new entity, venous dissecting aneurysm.

Key Words: venous aneurysm, venous dissection, medial vascular channels.

Introduction

Venous aneurysm particularly of upper extremity is an extremely rare but established entity [1]. We presented an extremely rare case of a 38-year-old female's venous aneurysm of left cephalic vein with unique histopathological features, displaying variably thinned medial wall with focal, markedly reduced or absent smooth muscle cells and elastic fibers, most likely leading to the venous dissection with an intimal tear and many medial blood-filled vascular channels. We herein propose that our venous dissection-like findings would be a new feature especially from the clinicopathological viewpoints and might be considered in the classification of venous aneurysm.

Case Report

The patient, a 38-year-old female, complained of an extensive, painful and pigmented purpura with some palpable and compressible nodules on the lateral side of her left upper arm. Retrospectively, she showed sudden onset of transient chest pain and dyspnea approximately 2 months before the surgery. Except for that, she had an unremarkable medical history, such as any recent/previous injury or intravenous drug injection to her extremities. The laboratory data were within normal limits. A color

Doppler ultrasound sonography confirmed the presence of two small fusiform venous aneurysms filled with thrombi, in a background of normal venous waveform without any evidence of arteriovenous fistula. Since the patient requested surgical resection of the aneurysms, a longitudinal skipped incision was performed below the aneurysmal nodules and along the course of left cephalic vein (Figure 1A). Multiple cystic and bead-like appearing venous aneurysms were present, measuring $12 \times 11 \times 10$ mm or $16 \times 14 \times 14$ mm, respectively (Figure 1A).

On the scanning magnification of one aneurysm (Figure 1B), its transverse section showed that a localized and abnormally dilated venous lumen was filled with mixed stratified thrombi, surrounded by variably thinned medial wall with focal, markedly reduced or absent medial smooth muscle layer and elastic fibers. Elastica van Gieson (EVG) staining clearly revealed complete loss and focal dissection of medial to adventitial elastic fibers in the variably thinned aneurysmal wall (Figure 1C). Correspondingly, intimal fibromuscular thickening was partially recognized, accompanied by increased or decreased elastic fibers (Figure 1C) and associated with variously degenerated and fibrous change by the Masson's trichrome or alcian blue staining. Inflammatory foci were absent from the intima to adventitia, within our

thorough investigation. Microscopic findings demonstrated that there seemed to be an intimal tear extending into the deep layer of media of the aneurysmal vein, accompanied by the connection with the blood-filled vascular channels (Figure 1D). EVG and immunohistochemical smooth muscle actin (α -SMA) staining more clearly showed an elastic (Figure 1E) and fibromuscular (Figure 1F) tear in the medial wall, associated with reduced/absent and occasionally increased elastic fibers and smooth muscle cells (Figure 1C,E,F). Those dissecting vascular channels were lined by CD31-positive flattened endothelium. Overall, we finally made a diagnosis as venous aneurysm of the cephalic vein with possible venous dissection, and thus, these clinicopathological features would be proposed as a new entity; tentatively named venous dissecting aneurysm.

Discussion

To our knowledge, this is the second reported case of a dissecting aneurysm, following the first case occurring as a popliteal mass early in the course of pregnancy in a 32-year-old woman [2]. Despite that, venous dissecting aneurysm, presenting as brachial nodules covered by pigmented skin, has not been described elsewhere. That is a

potentially new clinicopathological variant of venous aneurysm, displaying a localized and abnormally dilated venous lumen coexisted with an intimal tear extending into the medial wall, accompanied characteristically by the close connection with the blood-filled vascular channels. It has been reported that venous aneurysms involving the formation of thrombi in the neck, upper extremities and thorax clinically results in the patient's acute pulmonary embolisms [2,3], most likely as in our case. Hence, although guidelines for the treatment of venous aneurysms have never been established, any surgical intervention should be the first-line therapy, especially but likely in case of dissection-induced local pain and/or cutaneous hemorrhage. Finally, according to our examinations together with several reference papers [1–3], the present venous aneurysm might be caused by degenerative changes in the wall at least in part, but not pregnancy, trauma, inflammation or congenital in origin.

Among the clinicopathological differential diagnoses of the current case, the most important one is a large varicose vein. In marked contrast to venous aneurysm, varicose veins of the lower, but not upper, extremities are very common disorders [4]. Furthermore, varicose veins histologically show variable intimal hyperplasia, fibrosis, and overall dilation together with characteristic vascular wall thickening [5]. Whereas,

histopathological features of venous aneurysms basically demonstrate variable wall thinning, as in the present case. In fact, within our thorough investigation, there have been no reports regarding dissecting varicose veins. In this context, we can resolve the distinction between these two entities easily.

In conclusion, we presented an extremely rare case of a 38-year-old female's venous aneurysm of left cephalic vein with unique histopathological features, displaying variably thinned medial wall with focal, markedly reduced or absent smooth muscle cells and elastic fibers, most likely leading to the venous dissection with an intimal tear and many medial blood-filled vascular channels. We propose that those venous dissection-like findings would be a new feature especially from the clinicopathological viewpoints and might be considered in the classification of venous aneurysm. Further collecting cases are needed to validate the presence and significance of venous dissecting aneurysm as a new histopathological entity. This short report could interest the scientific community, taken together with potentially specific findings of new entity, venous dissecting aneurysm.

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Figure 1

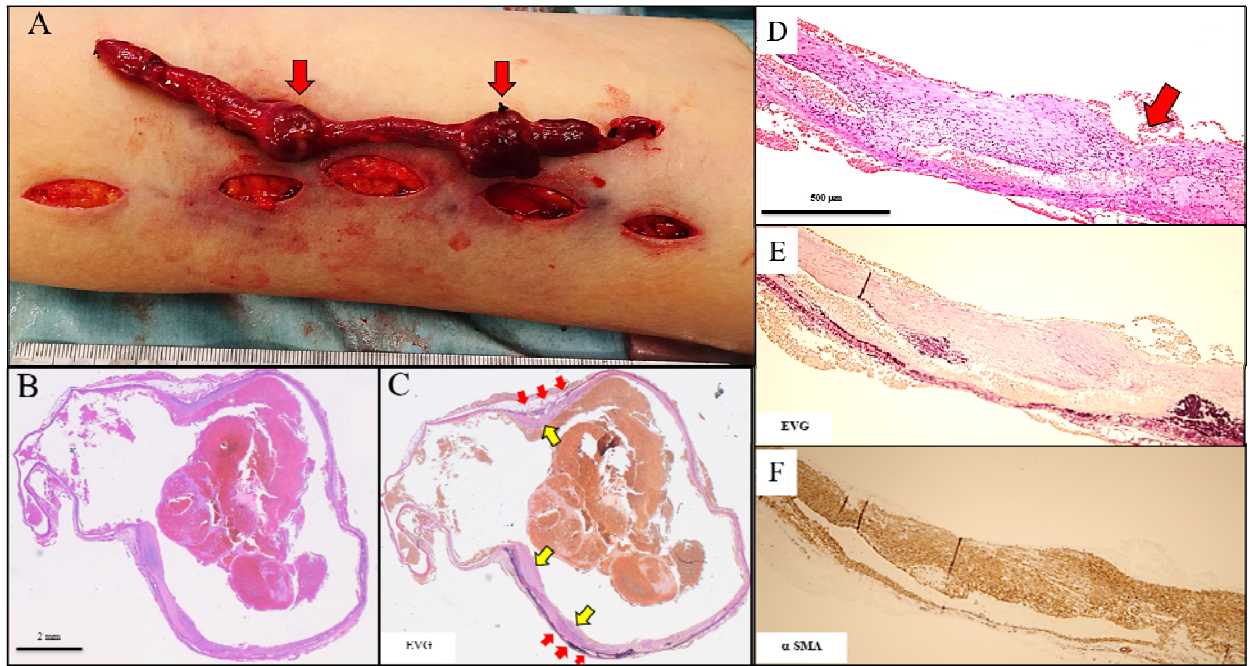


Figure Legends

Figure 1. Clinicopathological findings on venous aneurysm of the cephalic vein with possible venous focal dissection (venous dissecting aneurysm). (A) At surgery, a longitudinal skipped incision was performed below the aneurysmal nodules and along the course of left cephalic vein. Multiple cystic and bead-like appearing venous aneurysms were present, measuring $12 \times 11 \times 10$ mm or $16 \times 14 \times 14$ mm, respectively (red arrows). The skin revealed an extensive, pigmented purpura on the medial side of her left upper arm. (B) The transverse section of the aneurysm (H&E stain) showed that

a localized and abnormally dilated venous lumen was filled with mixed stratified thrombi, surrounded by variably thinned medial wall with focal, markedly reduced or absent medial smooth muscle layer and elastic fibers. Bar = 2 mm. (C) Elastica van Gieson (EVG) staining clearly revealed complete loss and focal dissection of medial to adventitial elastic fibers in the variably thinned aneurysmal wall (red arrows). Correspondingly, intimal fibromuscular thickening (yellow arrows) was partially recognized, accompanied by increased or decreased elastic fibers. (D) Microscopic findings (H&E stain) showed that there seemed to be an intimal tear (red arrow) extending into the deep layer of media of the aneurysmal vein, accompanied by the connection with the blood-filled vascular channels. Bar = 500 μ m. (E)(F) EVG (E) and immunohistochemical smooth muscle actin (α -SMA) staining (F) more clearly showed an elastic (E) and fibromuscular (F) tear in the medial wall, associated with reduced/absent and occasionally increased elastic fibers and smooth muscle cells.