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Palmar Digital Vein Thromboses: Their Different Expressions

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Key Words
Thrombosis of digital vein · Digital varicophlebitis · Mondor's phlebitis of the finger · Obliterating phlebitis · Antiphospholipid antibodies

Abstract
There have only been a few reports about thrombotic events in the volar digital veins. The observation of 2 such cases gives reason to discuss this rare symptomatology. Thrombosis of the digital veins can be categorised into three subtypes: the first shows clinical similarity to thrombotic events in veins or varicose veins of the limbs and may be related to a hypercoagulable state; the second group develops in a pre-existing normal vein or an acquired venous cavernoma and does not show clinical or histological signs of inflammation; the third category resembles Mondor's disease and may be named Mondor's phlebitis of the finger; histopathological examination has yet to be done. In any case of thrombosis of the volar digital veins, an inherited or acquired hypercoagulable state must be ruled out.

Introduction

65 years ago, Jadassohn [1] reported spontaneous thrombosis in small volar digital veins for the first time. He described his patient as 'psychologically unstable,... she didn't have varicose veins'. This means that Jadassohn tried to see this thrombotic event as being related to another underlying disease. He lacked our knowledge of the inherited and acquired hypercoagulable state. Therefore he interpreted his case as 'a new variant of tubercula dolorosa, which are of a very different nature (myoma, neuroma, glomangioma).

Until today only 11 cases with this rare thrombotic complication have been reported; none of them have appeared in the English dermatological literature.

Case Reports

Case 1

A 34-year-old mother and housewife noticed the growth of 5 small nodules on the volar aspects of different fingers within 1 year. The biggest of these nodules was meant to be removed because of the pain it caused her when pressure was applied. Her personal medical history was absolutely uneventful. She gave birth to 2 healthy children in 1992 and 1996 after undergoing normal pregnancies. She is a non-smoker with a BMI of 23.4. Since March 1998, the patient has had an intrauterine device (IUD) containing 52 mg of levonorgestrel (Mirena®). Physical examination revealed 2 slightly bluish nodules on the palmar aspect of the proximal interphalangeal (PIP) and just distal to the distal interphalangeal (DIP) joint of the left ring finger measuring 3 and 5 mm in diameter, respectively. Two further nodules were found on the palmar surface of the PIP joint of the right index finger (fig. 1) and the last over the PIP joint of the right middle finger. The patient showed a somewhat pronounced dorsal hyperextension in her metacarpophalangeal and interphalangeal joints (fig. 2), but there were no other signs which would indicate the possibility of an inherited connective tissue disease such as Ehlers-Danlos syndrome, Marfan syndrome or hypermobility syndrome. On histological examination, a cystic formation lined by a single layer of endothelial cells was found with fresh bleeding beside an old organizing thrombus and a normal vein wall (fig. 3). There was no inflammatory infiltrate. The finding was compatible with a partly thrombosed acquired venous haemangioma or venous cavernoma. Laboratory tests showed a normal haematogram. The search for an acquired or inherited hypercoagulable state revealed (with 50 U/ml; normal <20 U/ml) an isolated but marked increase in IgM antiphospholipid antibodies (APA). There were no IgG/IgA APA.

Case 2

A 49-year-old physician relates two episodes of painful swelling and hardening of a digital vein palmar and radial over the PIP joint of the left middle finger (fig. 4). The episodes occurred while doing intense gardening. The vein also became bent and somewhat transparent. The symptomatology disappeared within a few hours without treatment. Active massage accelerated the healing process. No histological examination has been undertaken.

Discussion

In 1936, Jadassohn [1] described for the first time the spontaneous development of thrombosis in digital veins: a 33-year-old

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woman had noted the presence of 2 small nodules in her left hand 6 months previously while using a cooking pan. These occasionally disturbed her as she worked on different chores in the kitchen and garden. Jadassohn felt 2 invisible nodules, about the size of a lentil, one of them on the palmar aspect of the left index finger just distal to the DIP joint and the second on the palmar surface of the DIP joint of the left fifth finger. There was no marked venous drawing and no varicosities on the palmar aspect of the hands and fingers. Histological examination revealed 'a vein containing an organizing thrombus'.

It was Jadassohn once again who presented a 46-year-old woman with the same symptomatology in 1959 [2].

In 1965, Martorell [3] observed 2 patients, who complained of painful hardening and swelling of an otherwise hardly visible vein on the palmar aspect of one finger. Martorell and his patients presumed that the symptomatology was caused by heavy manual work. Since it disappeared within 2–3 days, no histological examination was made. These two episodes showed a resemblance to Mondor's disease.

In 1972, Wilensky and Hubbard [4] described a 27-year-old woman with moderately swollen fingers on both hands, which were very tender to both pressure and joint motion. The symptomatology on the fingers 'reminded somewhat of phlegmasia coerulea dolens' and was preceded by two episodes with blurred vision. Wilensky and Hubbard [4] presumed that it was a thrombosis of digital vessels caused by the intake of contraceptive pills. Physical examination and extensive laboratory tests did not reveal any further pathological results. After the oral contraceptive had been discontinued, the patient became asymptomatic within a few weeks. No histological examination was done.

In 1978, Lechner et al. [5] observed the only reported patient with recurrent thrombotic complications within digital *varicose* veins. She was a 63-year-old woman who simultaneously suffered from recurrent pulmonary embolism. The singular varicophlebitis lasted about 3 weeks and resolved spontaneously. Histological examination showed 'a massive dilated vein partly obliterated by an organising thrombus. The media is somewhat fragmentated and there are some histiocytes containing hemosiderin and extravasations of red cells around the vessel wall'. Further physical examination and laboratory tests did not reveal any explanation for this hypercoagulable state.

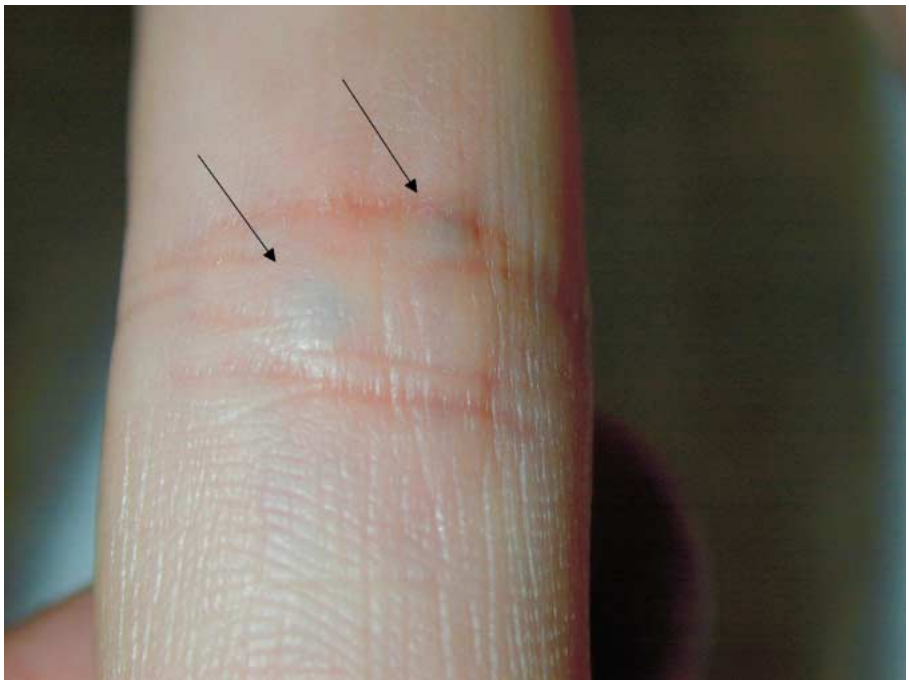

Fig. 1. Two slightly bluish nodules on the palmar aspect of the PIP joint (case 1).

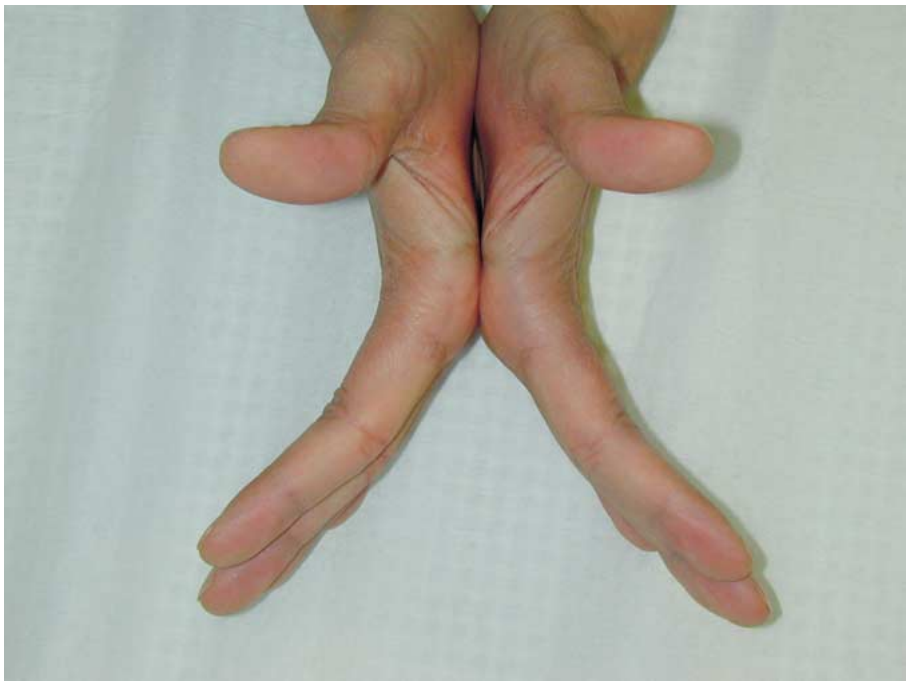

Fig. 2. Pronounced dorsal hyperextension in the metacarpophalangeal and interphalangeal joints (case 1).

Palmar and finger varicosities are not so rare. Clark et al. [6] find them in up to 52% in elderly people. Kutzner et al. [7] confirm that frequency, but Braga Da Silva and Foucher [8] describe 12 patients with palmar varicosities with an average age of only 41 years. Considering these results, it is astonishing that there have been no further reports in the literature telling us about *thrombotic* complications in palmar and finger varicosities. More reports concern spontaneous *haematomas* of the finger, also called the Achenbach syndrome [9], which is probably mechanically induced, preferring finger varicosities [10].

60 years after the first description of spontaneous thrombosis of palmar digital veins, Lanzetta and Morrison [11] reported another 5 women (aged 37–68 years, mean 53.4) in 1996 who showed the same history and symptomatology as the first case described by Jadassohn [1]. The small nodules, persisting over a period of a few months, were not painful, but according to the changed lifestyle women now lead, they complained of the 'inability to wear rings, drive the car and play golf'! Histological examination showed 'cystic soft tissue partly filled by blood and partly by gelatinous material consistent with a thrombus in a dilated vein'. Further information concerning coexistent diseases which could be responsible for an acquired hypercoagulable state or the intake of a hormonal medication are not mentioned. The authors presume that these superficial veins, protected only by a very thin and pliable skin, are more vulnerable to trivial, unrecognized trauma.

In my case, it is a 34-year-old woman. Her personal medical history is absolutely uneventful. Within 1 year she notes the growth of 5 small nodules in her hands. Histological examination reveals an elderly organizing thrombus within an acquired venous haemangioma or venous cavernoma [12]. The same histological patterns showed the cases described by Lanzetta and Morrison [11]. There is no inflammatory infiltrate which could resemble periarteritis nodosa or thrombo-angiitis obliterans.

Since March 1998, the patient wears an IUD containing 52 mg levonorgestrel (Mirena). Hormonal contraceptives increase the natural incidence of venous thrombosis of 1–2/10,000 women per year 3- to 4-fold [13]. The IUD of the patient releases 20 μg at the beginning, 11 μg after 5 years, with a mean of 14 μg levonorgestrel per day. The systemically available dose lies between 150 and 200 pg/ml, which means it is 20- to 100-fold

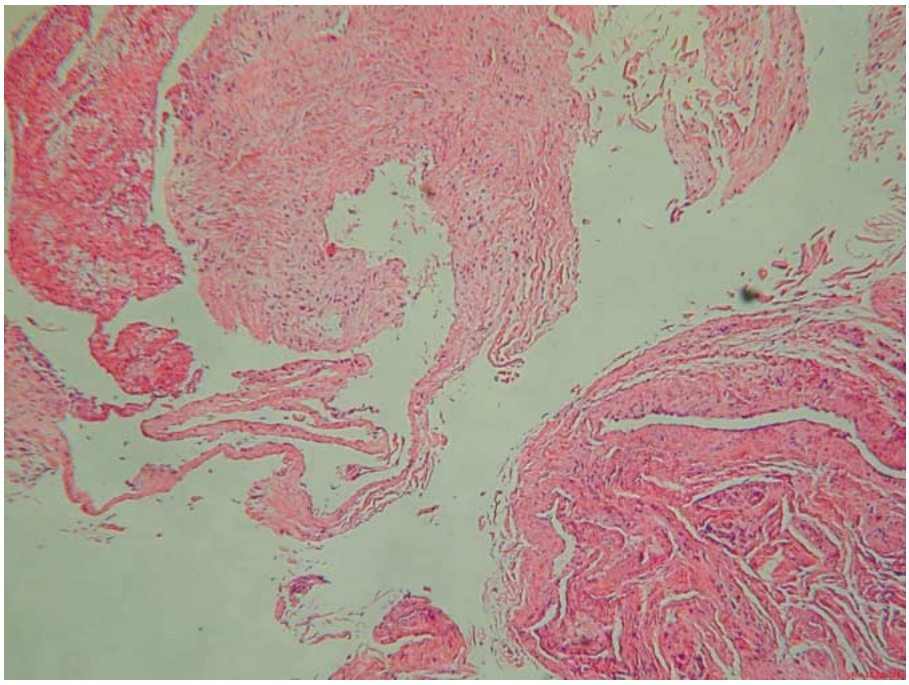

Fig. 3. On histological examination a cystic formation lined by a single layer of endothelial cells is found with fresh bleeding and elderly organizing thrombus beside thick- and thin-walled vessels. HE. ×40.

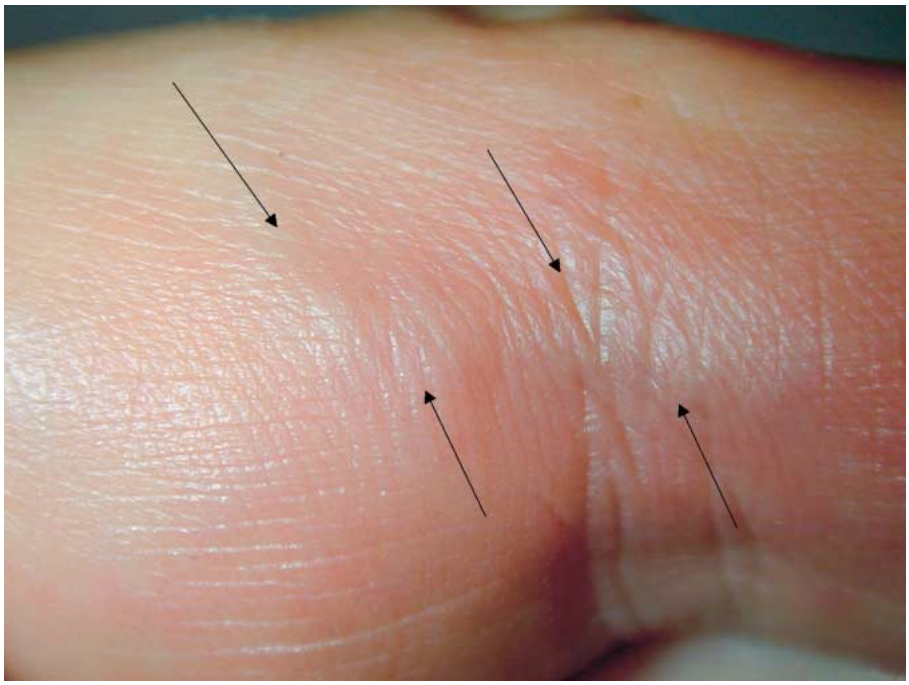

Fig. 4. Bent and swollen vein palmar and radial over the PIP joint of the left middle finger (case 2).

lower in comparison with the systemically available dose of an oral contraceptive of the second generation [14]. It is therefore more likely that the anticonception is not responsible for this thrombotic event.

The patient has clearly elevated IgM APA. APA are strongly associated with thrombosis and appear to be the most common of the acquired blood protein defects causing thrombosis [15]. Venous thrombosis is more common than arterial thrombosis in patients with a hypercoagulable state associated with APA [16], but the prevalence of IgM APA in a healthy population lies between 5.6 and 9.4% [17, 18], and the personal history of the patient reported here does not tell us anything about other thromboembolic events, recurrent fetal loss or thrombocytopenia. Therefore it is not yet possible to conclude whether the increased IgM APA are responsible factors in the pathogenesis of this thrombotic event. Further history will show whether digital vein thrombosis can be interpreted as an early sign of the antiphospholipid syndrome.

Another factor which could be responsible for the development of these thromboses is the fact that the patient here has a hyperextension of her finger joints.

The second case is comparable with the 2 patients reported by Martorell [3]: the symptomatology shows resemblance to Mondor's disease or, even more so, the so-called *Mondor's phlebitis of the penis* [19], which can sometimes be a real thrombotic event [20] but more often is an obliterating endophlebitis [21]. Until today, a histological examination of this symptomatology is still missing, and, thus, the question 'is it a lymphangitis or a phlebitis?' – which existed for years concerning the disease elsewhere also called *Hoffmann's lymphangitis of the penis* [22] – still remains unresolved.

Conclusion

Considering the 13 cases presented here, thrombosis of the digital veins can be classified into the following three categories:

The patients described by Wilensky and Hubbard [4] and Lechner et al. [5] show clinical similarity to thrombotic events in veins or varicose veins of the limbs. Corresponding to them are the appearance of inflammation, which are rubor, dolor, calor and tumor. They are presumably related to an acquired hypercoagulable state. Duration: a few weeks.

The second category concerns the cases observed by Jadassohn [1], Jadassohn and Golay [2], Lanzetta and Morrison [11] and the female patient reported here: within a normal vein or an acquired venous cavernoma, a thrombosis develops without clinical or histological signs of inflammation. The thrombosis persists for several months. It is supposed that trivial, unrecognized trauma causes it.

The third category, fitting the patients described by Martorell [3] and the male patient reported here, resembles Mondor's disease and may be named Mondor's phlebitis of the finger. It is presumed that the symptomatology is caused by heavy manual work. Histological examination does not exist. It is more presumably an obliterating endophlebitis than a real thrombosis. Duration: hours to days.

The diagnosis of thrombosis of the volar digital veins is made by the typical clinical aspect. Usually a histological examination is not necessary, but an exact investigation of the personal medical history, a thorough physical examination and in addition laboratory tests must be done, to confirm or rule out an acquired or inherited hypercoagulable state.

The therapy depends on the results of the investigations and the symptomatology: treatment of a causative underlying disease, withdrawal of causative medicaments (oral contraceptives), systemically given or locally applied antiphlogistic drugs (if necessary) and sometimes excision of a painful thrombotic nodule.

References

1 Jadassohn W: Ein Fall von Thrombosen in den Fingervenen. Schweiz Med Wochenschr 1936; 66:549.
2 Jadassohn W, Golay M: De la thrombose des veines des doigts. Minerva Dermatol 1959;34: 268.
3 Martorell A: Thrombosis venosa digital. Angiologia 1965;17:191–192.
4 Wilensky RJ, Hubbard TB Jr: Thrombosis of the digital vessels secondary to oral contraceptives. Am J Obstet Gynecol 1972;113:1137–1138.
5 Lechner W, Metz J, Viehwege G: Palmar- und Digitalvarikosis mit rezidivierenden Thrombophlebitiden. Dermatologica 1979;158:361–367.
6 Clark ANG, Melcher DH, Hall-Smith P: Palmar and finger varicosities of the aged. Br J Dermatol 1974;91:305–314.
7 Kutzner H, Hamann D, Wolff HH: Finger varicose veins of the aged. Hautarzt 1984;35:100–103.
8 Braga Da Silva J, Foucher G: Digital varices: 12 cases. Ann Chir Main Memb Super 1991; 10:319–323.
9 Hornemann M: Achenbach-Syndrom. Arch Dermatol Forsch 1972;244:603.
10 Stieler W, Heinze-Werlitz C: Paroxysmales Fingerhämatom (Achenbach-Syndrom). Hautarzt 1990;41:270–271.
11 Lanzetta M, Morrison WA: Spontaneous thrombosis of palmar digital veins. J Hand Surg (Br) 1996;21:410–412.
12 Hundeiker M: Cavernöse Angiome und benigne Neubildungen der Gefässwände; in Doerr W, Seifert G, Uehlinger E (eds): Spezielle pathologische Anatomie. Berlin, Springer, 1979, vol 7: Histopathologie der Haut, part 2, pp 328–329.
13 Bauersachs R, Kuhl H, Lindhoff-Last E, Ehrly AM: Risk of thrombosis with oral contraceptives: Value of a thrombophilia screening test. Vasa 1996;25:209–220.
14 Morant J (ed): Arzneimittelkompendium der Schweiz. Basel, Documed, 2001, pp 1619–1621.
15 Bick RL, Baker WF: Antiphospholipid syndrome and thrombosis. Semin Thromb Hemost 1999;25:333–350.
16 Provenzale JM, Ortel TL, Allen NB: Systemic thrombosis in patients with antiphospholipid antibodies: Lesion distribution and imaging findings. AJR Am J Roentgenol 1998;170:285–290.
17 Shi W, Krilis SA, Chong BH, Gordon S, Chesterman CN: Prevalence of lupus anticoagulant and anticardiolipin antibodies in a healthy population. Aust NZ J Med 1990;20:231–236.
18 Vila P, Hernandez MC, Lopez-Fernandez MF, Battle J: Prevalence, follow-up and clinical significance of the cardiolipin antibodies in normal subjects. Thromb Haemost 1994;72:209–213.
19 Tanii T, Hamada T, Asay Y, Yorifuji T: Mondor's phlebitis of the penis: A study with factor VIII related antigen. Acta Derm Venereol 1984;64:337–340.
20 Doi Y, Takeyama M, Matsui T, Fujioka H: A case of Mondor's disease of the penis. Hinyokika Kiyo 1988;34:1245–1248.
21 Braun-Falco O, Plewig G, Wolff HH: Erkrankungen von Glans penis and Präputium; in Braun-Falco O, Plewig G, Wolff HH (eds): Dermatologie und Venerologie, ed 4, vollständig überarbeitete und erweiterte Ausgabe. Berlin, Springer, 1996, p 1082.
22 Aragona P, Piazza R, Artibani W, Dante S: The so-called Hoffmann's lymphangitis of the penis: Is it a lymphangitis or a phlebitis? Int Urol Nephrol 1988;20:139–145.