

THE CHALLENGING OF DIAGNOSIS, TREATMENT, AND EDUCATION IN PATIENT WITH BUERGER'S DISEASE: A CASE REPORT FROM REMOTE AREA

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ABSTRACT

Background: Thromboangiitis obliterans (TAO) or Buerger's Disease is a chronic inflammatory disease. If not treated early and adequately, TAO can result in limb amputation. The study regarding TAO in women is rare and limited. Also how difficult it is to diagnose and educate patients with TAO, especially in remote areas

Case Presentation: A 38-year-old woman came to the emergency room with the chief complaint of pain and darkening in toes and hands 2 months ago, especially when it is cold. The patient did not come to the public health facility but was treated with traditional medicine according to local beliefs. History of smoking was denied. The localized status was found gangrene in toes and fingers, ulcer in toes, and oedema in legs. Allen's tests were positive. Dorsalis pedis arteries were palpable. ABI wasn't performed. Laboratory examination showed an increased liver function. Cardiomegaly, Lung oedema, and minimal right pleural effusion were shown in Chest X-ray. History taking, clinical finding, and laboratory finding suggested the patient to amputation but the patient refused it. The Patient was treated by antibiotic, analgetic, anticoagulant, and double. After 3 days of treatment, the patient was discharged and asked to return to the outpatient care for education about amputation.

Discussion: Most patients are aged 20-45 years. Men are more common than women. The etiology of TAO is unknown, but cigarette exposure is still associated with TAO. The role of hypercoagulability in the pathogenesis of TAO has been proposed. Elevated plasma homocysteine has been reported in patients with TAO. There was a strong link between the plasma Hcy concentration and Liver Fibrosis. We assessed the patient with TAO regarding suspect hypercoagulability affected by increased liver function. Difficult to diagnose due to patient denied smoking history and other imaging and laboratory tests weren't performed. Patients with TAO who have elevated homocysteine also have a higher rate of amputations. Education is a key role for understanding the concept of health. Countries with higher education

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levels are more likely to have better national health conditions. We can conclude that lower levels in tertiary education affects a country's health situation in terms of premature mortality. Conclusion: It is difficult to diagnose Buerger's disease or thromboangiitis obliterans because of the atypical complaints, minimal risk factors, and limited facilities for imaging and other laboratory tests. The difficulty of educating patients to get the best treatment is also a challenge for general practitioners, especially in remote areas

Keywords: Buerger's disease, thromboangiitis obliterans, woman, remote area, education level

INTRODUCTION

Thromboangiitis obliterans (TAO) or Buerger's Disease is a nonatherosclerotic, segmental, chronic inflammatory disease. TAO mainly affects the small and medium-sized arteries, veins and nerves of the arms and legs. Affected patients usually suffer from ischemic pain, ulcers, or gangrenous patches in the distal limbs. If not treated early and adequately, TAO can result in limb amputation (1). TAO is most prevalent in the Mediterranean, Middle East, and Asia. Men are more commonly affected than women. The annual incidence is 12.6 per 100,000 in the United States. In addition, there is a higher incidence in men than women (9:1), and the average age of presentation is 40-45 years (2). The information regarding TAO in women is rare and reported cases are limited. Also how difficult it is to diagnose and educate for management of patients with TAO, especially in remote areas which do not have sufficient facilities to perform imaging..

CASE PRESENTATION:

A 38-year-old woman came to the emergency room with the chief complaint of pain and darkening in toes and hands 2 months ago, especially when it is cold. Firstly, the patient complained of an ulcer on the toes then up to hands. Currently the

patient felt fever and jaundice. The patient did not come to the public health facility but was treated with traditional medicine according to local beliefs. The patient denied a history of chest pain, leg swelling, and any leg pain during walking. History of smoking, hypertension, diabetes mellitus, stroke, and other comorbidities were denied. As a teenager, the patient did not complain of any symptoms suggestive of an autoimmune disease such as malar rash, hair loss, and joint pain. History of taking drugs was denied. The patient had a history of thyroidectomy 1 month before the complaints appeared.

Physical examination showed an increase in respiration rate (28x/min) and normal in other vital signs. The localized status was found gangrene in toes and fingers, ulcer in toes, and oedema in legs. Other physical examinations including Allen's test were positive and dorsalis pedis arteries were palpable. ABI wasn't performed. Laboratory examination showed Leukosit 19.270 u/L, Hb 14.6 g/dL, Platelet 203.000 u/L, Blood glucose 72 mg/dL, Ureum 156 mg/dL, BUN 73 mg/dL, Creatinine serum 1.99 mg/dL, SGOT 113 U/L, SGPT 90 U/L, Albumin 2.7 g/dL, Cholesterol total 69 mg/dL, Triglycerida 73 mg/dL, HDL-C 8 mg/dL, LDL-C 52 mg/dL, and HbsAg non reactive. ECG showed Sinus Rhythm with 121 bpm

regular and no other abnormalities. Chest X-ray showed cardiomegaly suspect mitral heart disease with lung edema and minimal right pleural effusion.

History taking, clinical finding, and laboratory finding suggested the patient to amputation but the patient refused it. The Patient was treated by Ceftriaxone 1gr/12 hour IV, Ranitidin 50 mg/8 hour IV, Ketorolac 30mg/8 hour IV, loading heparin 5000 IU/IV/Syringe pump, maintenance heparin 1000 IU/hour/syringe pump, Cilostazole oral 100 mg/8 hour, Clopidogrel oral 75mg/24 hour, Drip farbion/24 hour. After 3 days of treatment, the patient was discharged and asked to return to the outpatient care for education about amputation.

DISCUSSION:

Thromboangiitis obliterans (Buerger disease) is a nonatherosclerotic, inflammatory vasculitis located in small-to-medium-sized blood vessels. The walls of the vessels are usually spared until a much later stage when the entire distal vessel fibrosis. Buerger's disease occurs most often in the extremities and digits (3).

Most patients with Buerger disease are aged 20 - 45 years. It does not occur in the pediatric or elderly population. It is more common in men with a male-to-female ratio of 3:1 (4).

The etiology of thromboangiitis obliterans is unknown, but use or exposure to tobacco is central to the initiation and progression of the disease. If the patient smokes, stopping completely is an essential first step of treatment (3). Despite its strong association with tobacco exposure, the general pathophysiology of the disease is still unknown (5). There has never been a well-documented case of Buerger's disease published in a patient proven not to be using tobacco or cannabis in some form (ie, confirmation of smoking cessation by measuring urine nicotine or cotinine levels and toxicology screen) (6).

The role of hypercoagulability in the pathogenesis of TAO has been proposed. Elevated plasma homocysteine (Hcy) has been reported in patients with TAO. This increase may be related to the high prevalence of heavy smoking or may be directly related to the disease itself (4). There was a strong link between the plasma Hcy concentration and Liver Fibrosis: the higher the level of plasma Hcy concentration, the more evident the degree of LF (7). In the present study, the levels of Hcy in liver disease patients were found to be highly significant. This could be due to the reason that the liver is the center for methionine and Hcy metabolism. So, any disturbance in liver function can affect these metabolisms. Defect in the

metabolism leads to the increased serum Hcy levels (8).

Following Olin's diagnosis criteria for thromboangiitis obliterans such as: (1) Typically between 20 and 40 years old and male, although recently females have been diagnosed. (2) Current (or recent) history of tobacco use. (3) Presence of distal extremity ischemia (indicated by claudication, pain at rest, ischemic ulcers or gangrene) documented by noninvasive vascular testing such as ultrasound. Exclusion of other autoimmune diseases, hypercoagulable states, and diabetes mellitus by laboratory tests. (4) Exclusion of a proximal source of emboli by echocardiography and arteriography. (5) Consistent arteriographic findings in the clinically involved and non-involved limbs (3). We assessed the patient with buerger's disease or thromboangiitis obliterans regarding suspect hypercoagulability affected by increased liver function. Although difficult for us to diagnose thromboangiitis obliterans due to our patient not having a history of smoking and our hospital does not have sufficient facilities to perform imaging and other laboratory tests.

TAO mainly affects young patients who present ischemic ulcers, rest pain, claudication, coldness of extremities, but also migratory thrombophlebitis and

Raynaud phenomenon. Distal arterial occlusions can ultimately lead to limb amputation. In multivariate analysis, limb infection at diagnosis (HR, 12.1; 95% CI, 3.5–42.1; $P < 0.001$) was associated with a higher rate of amputation (extremity ischemia, ischemic ulcer/necrosis, limb infection), and arthralgia were the variables used in the multivariable model) (5). Patients with TAO who have elevated homocysteine may also have a higher rate of amputations than those with normal homocysteine levels (4). The effectiveness of other treatments including vasodilating or anti-clotting drugs (3).

Countries with higher education levels are more likely to have better national health conditions. Among the adult education levels, tertiary education is the most critical indicator influencing healthcare in terms of infant mortality, life expectancy, child vaccination rates, and enrollment rates. We can conclude that lower levels in tertiary education adversely affect a country's health situation in terms of premature

mortality. Understanding the health benefits of education is thus integral to reducing health disparities and improving the well-being of 21st century populations (9,10).

Less educated adults report worse general health, more chronic conditions, and more functional limitations and disability. Objective measures of health, such as biological risk levels, are similarly correlated with educational attainment, showing that the gradient is not a function of differential reporting or knowledge (10). The patient was suggested to amputate her toes considering that there is gangrene and signs of infection such as ulcer and increased leukocytes on lab results but the patient refused because she didn't care about the risk of complications that would occur and continued the traditional medicine according to local beliefs. It shows that education is a key role for understanding the concept of health.

CONCLUSION:

It is difficult to diagnose Buerger's disease or thromboangiitis obliterans because of the atypical complaints, minimal risk factors, and limited facilities for imaging and other laboratory tests. The difficulty of educating patients to get the best treatment is also a challenge for general practitioners, especially in remote areas.

REFERENCES

- Published in July 2024, included in Vol.1 (2024) : LHSJ July 2024 (10-16)
1. Zheng, Jie-Fu, et al. "The incidence and prevalence of thromboangiitis obliterans in Taiwan: A nationwide, population-based analysis of data collected from 2002 to 2011." *Clinics*, vol. 71, no. 7, 2016, pp. 399–403, [https://doi.org/10.6061/clinics/2016\(07\)08](https://doi.org/10.6061/clinics/2016(07)08).
 2. Seebald, Jessica, and Lyndon Gritters. "Thromboangiitis obliterans (Buerger disease)." *Radiology Case Reports*, vol. 10, no. 3, 2015, pp. 9–11, <https://doi.org/10.1016/j.radcr.2015.06.003>.
 3. Arkkila, Perttu ET. "Thromboangiitis obliterans (Buerger's disease)." *Orphanet Journal of Rare Diseases*, vol. 1, no. 1, 2006, <https://doi.org/10.1186/1750-1172-1-14>.
 4. Vijayakumar, Abhishek, et al. "Thromboangiitis obliterans (Buerger's disease)—current practices." *International Journal of Inflammation*, vol. 2013, 2013, pp. 1–9, <https://doi.org/10.1155/2013/156905>.
 5. Olin, Jeffrey W. "Thromboangiitis obliterans: 110 years old and little progress made." *Journal of the American Heart Association*, vol. 7, no. 23, 2018, <https://doi.org/10.1161/jaha.118.011214>.

6. Le Joncour, Alexandre, et al. “Long-term outcome and prognostic factors of complications in thromboangiitis obliterans (Buerger’s disease): A Multicenter Study of 224 patients.” *Journal of the American Heart Association*, vol. 7, no. 23, 2018, <https://doi.org/10.1161/jaha.118.010677>.
7. Lv, Dan, et al. “Plasma levels of homocysteine is associated with liver fibrosis in health check-up population.” *International Journal of General Medicine*, Volume 14, 2021, pp. 5175–5181, <https://doi.org/10.2147/ijgm.s329863>.
8. Kaur, Harsharan, et al. “Comparison of homocysteine levels in various liver diseases.” *JOURNAL OF CLINICAL AND DIAGNOSTIC RESEARCH*, vol. 13, no. 11, Nov. 2019, pp. BC01–BC03, <https://doi.org/10.7860/jcdr/2019/42661.13311>.
9. Raghupathi, Viju, and Wullianallur Raghupathi. “The influence of education on health: An empirical assessment of OECD countries for the period 1995–2015.” *Archives of Public Health*, vol. 78, no. 1, 6 Apr. 2020, <https://doi.org/10.1186/s13690-020-00402-5>.
10. Zajacova, Anna, and Elizabeth M. Lawrence. “The relationship between education and health: Reducing disparities through a contextual approach.” *Annual Review of Public Health*, vol. 39, no. 1, 1 Apr. 2018, pp. 273–289, <https://doi.org/10.1146/annurev-publhealth-031816-044628>