CASE REPORT

Acute idiopathic blue fingers: a young man with Achenbach's syndrome

Hidemi Takeuchi, Haruhito Adam Uchida, Yuka Okuyama, Jun Wada

Department of Nephrology, Rheumatology, Endocrinology and Metabolism, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama, Japan

Correspondence to
Dr Haruhito Adam Uchida,
hauchida@okayama-u.ac.jp

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SUMMARY

We report a case of a 20-year-old man presenting with acute painful blue fingers. All physical findings, including an Allen test, were normal, and systematic symptoms frequently seen in collagen diseases were absent.

Although we performed a wide variety of investigations including medical imaging, no specific abnormal findings were observed. Skin biopsy pathology was an important reference. The patient's symptoms gradually improved and were completely resolved without specific treatment. Based on the clinical presentation and course, we gave a diagnosis of Achenbach's syndrome, developed in a young male. Achenbach's syndrome is rare, but still may be encountered in clinical practice. The symptoms can be startling to the patient, eliciting fear of something terrible when, in fact, the syndrome is relatively benign and has a good prognosis. Recognising this disease quickly after presentation helps to eliminate the anxiety of the patient, as well as reducing excessively invasive investigations. We present a case report to enlighten Achenbach's syndrome.

BACKGROUND

Achenbach's syndrome is a disease that was proposed by German physician Achenbach, in 1958.¹ Patients have haematomas and purpuras on a finger or the palm, accompanied by abnormal sensations such as pain, numbness and coldness, without any apparent causes. These symptoms can last several days to several weeks, disappearing naturally. Occasionally, a pallor change is seen and a swollen feeling can cause temporary movement disorder due to compression of the surrounding tissue.

There is surprisingly little known about this disease, although it can be commonly encountered in daily practice. It has been alternately reported as 'Acute blue finger',² 'Non-ischaemic blue finger',³ 'Spontaneous blue finger syndrome'⁴ or 'Neglected syndrome'⁵ in various countries by several authors.

Typically, middle-aged women develop it in their index finger or middle finger. Although symptoms appear repeatedly, the condition is not accompanied by significant complications, including bleeding.

Achenbach's syndrome is a benign disease with a good prognosis. Therefore, it is important to be aware of this syndrome to avoid stress causing, excessive and invasive procedures in patients. Recognising Achenbach's syndrome allows doctors to manage patients who present with acute blue fingers, ruling out other more serious diagnoses.

We report a rare case of Achenbach's syndrome in a young man.

CASE PRESENTATION

The patient, a 20-year-old man, presented in November with sudden pain, cyanosis and a cold feeling on the palmar side of the apex-to-base left thumb and index finger, without particular causative triggers. Initially, he had visited an orthopaedist and rheumatologist after symptoms persisted, however, a cause was not found. He was then introduced to our hospital the following month.

He was a non-smoker and did not consume alcohol, and had neither an abnormal medical nor family history. He was a member of the field track club at the University and had recently begun training with barbells.

On examination, the apices of his left thumb and index finger had a blue tinge (figure 1), and were tender and cold to the touch. A small ulcer had formed on the index fingertip. An Allen's test showed normal patency and there was no difference in blood pressure between the right and left upper limbs. Both right and left brachial, radial and ulnar arteries were well palpable. Raynaud's phenomenon, fever, joint pain, muscle pain and other systemic symptoms were not observed.

INVESTIGATIONS

Laboratory studies (table 1) showed no abnormalities in platelet count, coagulation, autoantibody, such as antinuclear antibody, anti-phospholipid antibody, etc.

Ankle Brachial Pressure Index and Pulse Wave Velocity (ABI/PWV) (figure 2) were normal. Skin Perfusion Pressure (SPP) (figure 3) was 46 mm Hg

Figure 1 The blue colour change on the apices of the left thumb and index finger.

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Table 1 Laboratory studies

WBC	5840	/ μ L	TP	7.7	g/dL	CRP	0.03	mg/dL
Ne	60.9	%	Alb	5.0	g/dL	CH50	42	U/mL
Ly	27.7	%	AST	40	IU/L	C3	108.3	mg/dL
Mon	5.6	%	ALT	35	IU/L	C4	22.8	mg/dL
Eos	5.2	%	γ -GTP	17	IU/L	IgG	1236.7	mg/dL
Bas	0.6	%	LDH	298	IU/L	IgA	238.5	mg/dL
RBC	531	$\times 10^4$ / μ L	CK	1698	IU/L	IgM	101.7	mg/dL
Hb	16.4	g/dL	Na	138	mEq/L	ANA	0.14	times
Ht	47.9	%	K	4.9	mEq/L	Anti-RNP antibody	0.67	U/mL
PLT	385	$\times 10^4$ / μ L	Cl	101	mEq/L	Anti-SS-A antibody	<5.0	IU/mL
PT-INR	0.94		Ca	10.2	mg/dL	Anti-SS-B antibody	<5.0	IU/mL
APTT	28.1	sec	UN	14.9	mg/dL	Anti-Scl-70 antibody	<5.0	IU/mL
Fibg	232	mg/dL	Cre	0.84	mg/dL	Anti-Jo-1 antibody	<5.0	U/mL
FDP	<2.5	μ g/mL	UA	4.7	mg/dL	Cardiolipin antibody	2.38	U/mL
D-dimer	<0.5	μ g/mL	T.Cho	194	mg/dL	Rheumatoid factor	<5.0	IU/mL
			TG	225	mg/dL			
PPG	98	mg/dL	HDL-C	63	mg/dL			
HbA1c	5.5	%	LDL-C	117	mg/dL			

Alb, albumin; ALT, alanine transaminase; ANA, antinuclear antibody; APTT, activated partial thromboplastin time; AST, aspartate transaminase; CK, creatine kinase; Cre, creatinine; CRP, C-reactive protein; FDP, fibrinogen degradation products; Fibg, fibrinogen; Hb, hemoglobin; HbA1c, hemoglobin A1c; HDL-C, high-density lipoprotein cholesterol; Ht, hematocrit; LDH, lactate dehydrogenase; LDL-C, low-density lipoprotein cholesterol; PLT, platelet; PPG, postprandial glucose; PT-INR, international normalized ratio of prothrombin time; γ -GTP, γ -glutamyl transpeptidase; RBC, ; RNP, ribonucleoprotein; T. Cho, total cholesterol; TG, triglyceride; TP, total protein; UA, uric acid; UN, urea nitrogen; WBC, white blood cell.

Figure 2 Ankle Brachial Pressure Index and Pulse Wave Velocity.

Figure 3 Skin Perfusion Pressure.

on the palmar side and 56 mm Hg on the dorsal side of the left hand at the base of the index finger.

On contrast-enhanced CT (figure 4), no stenosis of the aortic arch, nor of the subclavian, axillary, brachial, radial and ulnar arteries was observed. Relatively normal blood flow to the peripheral fingers and palmar arch was confirmed, and no other abnormalities in bone, organs and soft tissue were found.

Skin biopsy pathology images are shown in figure 5. Hyperkeratosis and parakeratosis around a small ulcer and vascularisation changes were observed. Other blood vessels were normal. A certain leakage of red blood cells below the dermis layer was found. Leucocytes had infiltrated the surrounding ulcer. No leukocytoclastic vasculitis, autoimmune vasculitis and thrombosis were observed.

DIFFERENTIAL DIAGNOSIS

We considered it important to check for any abnormalities in a complete blood count and fibrinolysis-coagulation system, to confirm the absence of rheumatic diseases that cause vasculitis or Raynaud's syndrome. We also wanted to rule out thrombosis, severe stenosis or an extravascular arterial obstruction that could cause ischaemia, as well as complications due to cholesterol crystal embolism (CCE).

First, there was no increase in red blood cells and no abnormalities in platelet count or the coagulation-fibrinolysis system, including d-dimer and FDP. Thus, we could exclude polycythaemia, thrombocytopenia and fibrinolysis-coagulation disorder.

Second, autoantibody findings and inflammation negated the possibility of rheumatic diseases, such as small vessel vasculitis or Raynaud's syndrome. Normal levels of complement and globulin were observed. Gross physical examination detected neither rash, scleroderma symptoms, arthritis, muscle pain, serositis nor abnormalities of the conjunctiva and mucous membranes. The patient had neither peripheral nor central nervous system disorders, nor gastrointestinal or respiratory symptoms. Furthermore, the focal blue colour change of the fingers did not match Raynaud's syndrome, where it is generally found on both sides of the hands and feet, and influenced by time of day and temperature. Buerger's disease was excluded due to lack of symptoms involving the feet and no history of active or passive smoking. The patient's testimony negated vibration-induced or cold-induced injury.

Subsequently, CT imaging revealed no evidence of large and middle vessel vasculitis, including Takayasu's arteritis and giant cell arteritis, thoracic outlet syndrome, extra vessel obstruction or any other atherosclerotic stenosis.

Finally, in skin biopsy pathology, we found no cholesterol crystal and no fibrin embolism in the vessels (which often accompanies CCE). In addition, we did not find vasculitis and thrombotic occlusion.

Given these observations, we could rule out other diseases and identify this as an episode of Achenbach's syndrome, albeit in an uncharacteristic patient demographic.

TREATMENT

At first, we considered the patient's symptom to be caused by a blood flow disorder, such as micro-thrombus or vasospasm. We therefore prescribed sarpogrelate hydrochloride, which

Figure 4 Contrast-enhanced CT.

Figure 5 Skin biopsy pathology image (H&E-stained).

selectively antagonises both, platelets and vascular serotonin receptors (5-HT₂), and inhibits the aggregation of platelets and the contraction of vessels. This treatment is expected to improve various ischaemic symptoms such as ulcer, pain and cold sensation. Since the patient's symptoms gradually subsided in the 1-month follow-up, we stopped the medication.

OUTCOME AND FOLLOW-UP

To confirm the therapeutic effect and to assess the patient's condition, we invited him to visit after 1 month, at which time his symptoms, such as the feelings of pain and coldness, had almost completely improved. However, the purpuras and a small ulcer on the finger had not disappeared; we then took a skin biopsy from his finger, to differentiate between Achenbach's syndrome and other collagen diseases. We observed him every 2 weeks to check the scar and to confirm his symptoms as Achenbach's syndrome. The finger colour and pain steadily normalised at each

subsequent visit. Finally, all symptoms disappeared within 2 months from the initial visit. At his last visit, we suggested a revisit if recurrent episodes occurred, however, he has, fortunately, not had to visit our office again.

DISCUSSION

This is a rare case of Achenbach's syndrome developing in a young man.

Achenbach's syndrome is diagnosed by the presence of characteristic episodes and exclusion of other diseases, shown in table 2, based on physical findings, imaging examinations and haematological tests.

As our patient was a young man, it was not a typical case. However, his course and symptoms were compatible with Achenbach's syndrome, and we could completely exclude other diseases. Consequently, we concluded he suffered from Achenbach's syndrome.

Typically, Achenbach's syndrome occurs in middle-aged women, usually involving the index finger and middle finger without particular contributing factors.^{2 5 6} The previous literature has demonstrated that symptoms often appear on the palmar side of the left hand, starting with the index finger, then middle and so on, in descending order of frequency. The disease is most likely to appear in the mid-joints and based-joints.^{2 5 6} In rare instances, it has affected the thumb,^{2 7} as well as the index or middle finger, similar to the present case. It has been reported that Achenbach's syndrome is diagnosed most often in middle-aged women, and that the average age is 55–57 years (range: 19–88 years).^{2 8 9} The ratio of women to men affected by the disease is roughly 1:2~7. Our case is unique in that the patient was both young and male, two atypical variables for this particular affliction.

Although many patients develop this disease, without any identifiable cause, there are several reports suggesting it to be induced by external force.⁸ A distribution of blue colour change on the opposing thumb-index finger, as observed in our case, can suggest an association with external force. Since this patient was training with barbells, it may be assumed that the act of gripping the barbell was enough of a trigger. Because symptoms generally occur in the left hand, which is non-dominant for

Table 2 Example of differential diagnosis	
Anatomic location	Pathologic entity
Aortic arch	Takayasu's arteritis Giant cell arteritis Aneurysmal disease producing emboli
Subclavian and axillary artery	Thoracic outlet syndrome Trauma Atherosclerosis
Brachial, radial, ulnar arteries and palmar arch	Collagen vascular disease Atherosclerosis Buerger disease Trauma Ulnar artery thrombosis
Digital arterioles	Raynaud syndrome Vasospasm Collagen vascular disease Microemboli Vibration-induced injury Cold injury Haematologic (polycythaemia)

most people, we have to consider that most patients may not notice their triggers.

The pathology of Achenbach's syndrome has rarely been displayed in previous reports.^{8 10} In our skin biopsy specimen, we observed hyperkeratosis and parakeratosis around a small ulcer demonstrating repair of skin, and a vascularisation change indicating a repair process of microarteries after microvascular disorder due to ischaemia or hypoxia. Most other vessels were normal. Indeed, these findings may be non-specific alterations that are observed in other diseases forming ischaemic ulcers, but these changes should be observed in Achenbach's syndrome. It is also important to confirm the absence of vascular changes that represent CCE or thromboembolism. In fact, we could not find any vessels representing these diseases. The leakage of red blood cells below the dermis layer was considered to be subcutaneous bleeding due to skin injury. Leucocytes that had integrated into the surrounding ulcer were assumed part of the natural wound healing process, rather than an indication of vasculitis. Skin biopsy pathology is an important reference to confirm regeneration after vascular disorders and to eliminate other vascular disease. Therefore, doctors may be allowed to biopsy, if symptoms are strong or a diagnosis is difficult.

Learning points

- ▶ We experienced a rare case of a young man with Achenbach's syndrome.
- ▶ While it is rare for patients with this syndrome to get their skin biopsied, skin biopsy pathology is one of the important references to confirm the regeneration after vascular disorders and to eliminate other vascular disease.
- ▶ While this disease in the acute phase exhibits significant clinical symptoms, the disease follows to regression in its natural course and leaves no sequelae.
- ▶ It is possible to eliminate the anxiety of patients, to reduce excessively invasive investigations and to prevent unneeded referrals, by recognising Achenbach's syndrome.

Due to highly visible and unusual symptoms, Achenbach's syndrome can cause much anxiety in the patient—for fear that it is something life-threatening. Indeed, Achenbach's syndrome is a benign disease that resolves—with neither short nor long term sequelae—and symptoms usually disappear within several days; most patients have no symptoms at 6-week follow-up and few patients get recurrent episodes in months or years.² If doctors are aware of this disease, they can reassure the patient of the favourable prognosis. Recognising this disease also leads to a reduction of excessively invasive investigations and unneeded referrals. We report to enlighten that Achenbach's syndrome, although rare, is possibly encountered in clinical practice and knowledgeable doctors should take the proper care to diagnose this benign disease.

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Contributors HT, YO and HAU carried out the actual medical care and data collection for this case. HT prepared the manuscript, and gathered the necessary materials and documents. HAU and JW gave advice on the case and on the manuscript.

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