Case Report

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Achenbach's hand and digital paroxysmal haematomas: a possible association with joint hypermobility syndrome in two sibling cases

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Summary

A pair of middle-aged sisters is presented who experienced episodes of spontaneous, relatively atraumatic, acute onset, painful sub-cutaneous bleeding into the digits of their hands over a number of preceding years. A literature search revealed they had Achenbach's syndrome, which is a benign and self-limiting condition that resolves completely within 2–4 days. However, a hitherto unreported association between Achenbach's syndrome and joint hypermobility with its abnormal collagen is made, along with a tentative causality hypothesis that perhaps in the presence of atypical connective tissue, the architecture and integrity of the peripheral capillary bed of the hand could be adversely affected and therefore predispose towards paroxysmal palm and/or finger bleeds.

Keywords

connective tissue disease, rheumatology, clinical, dermatology, general practice, family medicine

Introduction

The condition of either a palm or a proximal/middle finger phalange experiencing a sudden sharp pain, followed by volar, sub-cutaneous bleeding and blue-black discolouration with swelling and numbness that develops over a few minutes, was first described by Achenbach in 1957.[1]

Other cases have since been published,[2–9] involving restricted finger bending,[8] digital swelling in 60% of cases and paraesthesia and or itching in 25% of cases.[3]

As the haematoma enlarges, it spreads into the adjacent phalanges and the palm of the hand with ensuing pain relief,[2] but characteristically never into the fingernail or fingertip.[2,7]

The haematomas take a median of four days to disappear[3] and occur without the prolonged colour changes that are associated with an ecchymosis.[5]

The syndrome is characterised by recurrent episodes and while a third of cases arise from simple, every-day manual activities, most develop spontaneously and without any abnormal coagulation variables.[3,6]

In Britain, the female to male predilection is 6:1[7] with a median presentation age of 50 years,[3] but at present, there seem to be neither prevalence nor incidence data published for this condition, nor any aetiological explanations.

First case presentation

A non-smoking, 64-year-old female dental hygienist presented with a bruised and swollen right index finger, which arose spontaneously as a sudden pain on the volar-lateral aspect of the right index finger's proximal phalange. Within an hour, the bruising and swelling extended distally into the second phalange, but not into the fingertip (Figure 1(a)). It then progressed proximally into the palm of the hand (Figure 1(b)) with ensuing pain alleviation. Over the following three days, the discolouration, discomfort and swelling subsided and the finger's mobility normalised.

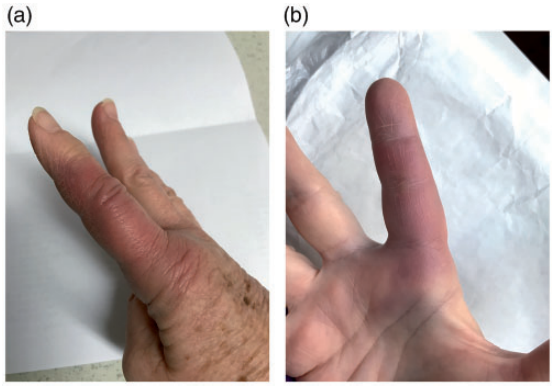
She gave a history of approximately a dozen similar episodes over the preceding few years, but never with thumb or little finger involvement.

On each occasion, her manual dexterity and clinical practice were diminished for several days.

Her medical history included a teenage onset of both migraine and paroxysmal supraventricular tachycardias; post-menopausal sub-clinical hypothyroidism and an affirmative score of one major and two minor Brighton criteria, which is indicative of probable joint hypermobility syndrome (Table 1), where joint hypermobility syndrome is diagnosed in the presence of two major Brighton criteria, one major and two minor criteria, four minor criteria, or two minor criteria when a first-degree relative is categorically affected.[10–12]

Of her minor Brighton criteria, she had suffered with intermittent joint pain in her right hip for three months (radiographic absence of trauma, neoplasia

Figure 1. (a) Subcutaneous bruising and oedema of the hygienist's right index finger, with sparing of the fingertip. (b) Bruising extending into the palm of the hygienist's hand.



or degenerative osteoarthritis) and she had had blepharoplasties to correct droopy eyelids, which had obstructed her vision.

A full blood count, clotting screening and a tourniquet capillary resistance test of Hess were all normal.

Second case presentation

The hygienist's 59 year-old non-smoking retired bank manager sister confirmed that her paroxysmal haematomas began in her mid-fifties, with the involvement of the fingers of both hands, but predominantly those in her dominant right hand. She reported only one episode in the palm of her hand (Figure 2(a)), but never any in either her thumb or little finger. She displayed similar signs and symptoms to her sister, and reported that (i) the bleeds are always associated

Table 1. The Brighton Major and Minor Criteria used to determine the joint hypermobility scores for the two siblings.

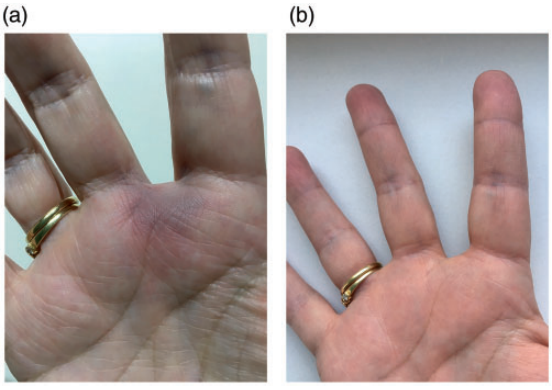
	Case 1's score	Case 2's score
Major Brighton criteria		
Arthralgia for >3 months in ≥4 joints	0	0
A Beighton score of 4 or more	1	1
Beighton scores		
Place hands flat on the floor keeping knees and legs straight?	0	1
Bend left elbow backwards?	0	1
Bend right elbow backwards?	0	1
Bend left knee backwards?	0	1
Bend right knee backwards	0	1
Bend left thumb back onto front of forearm?	1	1
Bend right thumb back onto front of forearm?	1	1
Bend left little finger at 90° towards the back of the hand?	1	1
Bend right little finger at 90° towards the back of the hand?	1	1
Totals of Beighton scores	4	9
Brighton minor criteria		
Beighton score 1–3	0	0
Arthralgia in 1–3 joints/back pain >3 months	1	0
Multiple joint single/Single joint multiple dislocations	0	0
≥3 episodes of tenosynovitis/bursitis	0	0

(continued)

Table 1. Continued.

	Case 1's score	Case 2's score
Marfanoid habitus	0	0
Thin, stretchy skin, striae, abnormal scarring	0	0
Droopy eyelids, myopia	1	1
Varicose veins, hernia, visceral prolapse	0	0
Totals of minor Brighton criteria	2	1

Figure 2. (a) Spontaneous haematoma in the palm of the sibling's hand. (b) Complete resolution of the palm haematoma 24 h later.



with activities such as carrying shopping or opening a stiff glass jar, (ii) the pain feels like a needle is being stuck into the finger and it looks like a blood vessel has burst, (iii) with volar bruising that spreads 3–4 mm beyond, but never with any swelling, and (iv) they always completely resolve within 24 h (Figure 2(b)). She responded affirmatively to one major and one minor Brighton criteria, but with a positive response to all of the Beighton scores, indicating a similar, probable involvement with joint hypermobility syndrome (Table 1).

Her medical history only involved two isolated grand mal episodes in the preceding six years, for which no cause could be found.

Discussion

The overlap between the signs and symptoms of Achenbach's syndrome and other circulatory diseases, such as Acute Limb Ischaemia and Raynaud's phenomenon can create a diagnostic challenge (Table 2).[3]

The application of either a circulatory disease algorithm,[3] or a sieve,[7] duplex sonography and or peripheral angiography may be used to exclude ischaemic conditions.[2]

However, given the characteristic presentations, the two- to four-day spontaneous regressions and the fact that Achenbach's syndrome has no association with serious bleeds into other organs,[2,3] the need for any invasive diagnostics is usually precluded,[2,5–7] just simply the delivery of some simple reassurance.[2,3]

Similar comorbidities that were present in the hygienist have been previously reported in other Achenbach patients, but no pathogenic connections have ever been established.[2]

For both cases, an association with joint hypermobility syndrome was apparent and a causal relationship is therefore tentatively hypothesised; namely, that the abnormal collagen could perhaps adversely affect the architecture and integrity of the peripheral capillary bed of the hand, thereby resulting in spontaneous paroxysmal palm or finger bleeds, in a similar conceptual way to how the abnormal extracellular matrix fibrillin in Marfan's syndrome predisposes these patients towards dissecting aortic aneurysms.

However, with just two cases demonstrating an association between Achenbach's syndrome and joint hypermobility, the credibility of a possible causal link can only be improved by the identification of more cases.

Conclusion

While Achenbach's syndrome is uncommon, the reported association with joint hypermobility syndrome that this case series illustrates should alert clinicians to become more vigilant in looking for comorbidity prevalence in other similar patients. As a consequence, in addition to haematological investigations, capillary fragility and coagulation blood tests for Achenbach syndrome patients, they should also be screened for joint hypermobility using the

Table 2. The similar and differentiating features between Achenbach's syndrome and two other major vascular disorders with digital presentations.

Demographics	Achenbach's syndrome	Acute limb ischaemia	Primary Raynaud's phenomenon
Affected anatomical sites	Volar aspect of a finger ± the palm	A complete limb.	Commonly, Fingers and thumbs, less commonly the toes and rarely the nose tip, ears, lips, tongue and nipples.
Seasonal/winter presentation	X	X	✓
Mean duration	2–4 days	Without intervention, indefinite	Typically, minutes, but can be several hours
Spontaneous resolution	✓	X	✓
Mean age of onset (years)	≥50	Any age, embolic/thrombotic event	15–30
British gender predilection	F > M 6:1	F = M 1:1	F > M 1.7:1.0
Signs			
Finger tips spared	✓	X	X
Discolouration	Blue	Initially marble white, progressing to light blue/purple reticular mottling (deoxygenated blood) and later, darker, coarser, mottling patterns (stagnant blood)	Classic Tricolour, from an initial white (vaso-spasm) to blue (deoxygenation) to red (reactive hyperaemia) sequence of colour changes
Oedema	✓	X	Post-episodic only, during recovery stage
Palpable peripheral pulse	✓	X	✓
Peripheral temperature	Normal	Reduced	Reduced
Symptoms			
Pain	✓	✓	Post-episodic during recovery stage
Paralysis	X	✓	X
Paraesthesia	✓	✓	✓

Brighton criteria questionnaire,[10–12] with referral to a rheumatologist or a specialist in connective tissue disorders, as appropriate. If joint hypermobility is found to co-exist more widely, this could stimulate further research into the role that abnormal collagen may have in compromising the architecture and integrity of the peripheral capillary bed of the hand.

Declarations

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References

1. Achenbach W. Paroxysmal hematoma of the hand. *Athena* 1957; 23: 187–189.
2. Thies K, Beschorner U, Noory E and Zeller T. Achenbach's syndrome revisited. *Vasa* 2012; 41: 366–370.
3. Kordzadeh A, Caine PL, Jonas A, et al. Is Achenbach's syndrome a surgical emergency? A systematic review. *Eur J Trauma Emerg Surg* 2016; 42: 439–443.
4. Robertson A, Liddington MI and Kay SP. Paroxysmal finger haematomas (Achenbach's syndrome) with angiographic abnormalities. *J Hand Surg (Brit and Eur Volume)* 2002; 27B: 391–393.
5. Deliss LJ and Wilson JN. Acute blue fingers in women. *J Bone Joint Surg* 1982; 64B: 458–459.
6. Dhaliwal K, Brewster CT and Pakeerathan S. Acute blue finger syndrome: a rare benign cause of acute digit discolouration. *Br Med J Case Rep* 2019; 12: e223365.
7. Khaira HS, Rittoo D, Vohra RK and Smith SRG. The non-ischaemic blue finger. *Ann R Coll Surg Engl* 2001; 83: 154–157.
8. Kämpfen S, Santa DD and Fusetti C. A painful blue thumb: a case of Achenbach's syndrome. *EJVES Extra* 2005; 10: 84–45.
9. Mishra P, Jain A, Sen S and Majumdar B. Recurrent bleeding and bruising over little finger: a diagnostic conundrum! *Indian J Dermatol* 2015; 60: 632–633.
10. Grahame R, Bird HA, Child A, et al. The revised (Brighton 1998) criteria for the diagnosis of benign joint hypermobility syndrome (BJHS). *J Rheumatol* 2000; 27: 1777–1779.
11. Beighton P, Solomon L and Soskolne CL. Articular mobility in an African population. *Ann Rheum Dis* 1973; 32: 413–418.
12. Tinkle BT, Bird HA, Grahame R, Lavallee M, Levy HP and Sillence D. The lack of clinical distinction between the hypermobility type of Ehlers-Danlos syndrome and the joint hypermobility syndrome (a.k.a. hypermobility syndrome). *Am J Med Genet Part A* 2009; 149A: 2368–2370.
13. Gagnier JJ, Kienle G, Altman DG, Moher D, Sox H and Riley D the CARE Group. The CARE guidelines: consensus-based clinical case reporting guideline development. *Glob Adv Health Med* 2013; 2: 38–43.