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An unusual case of superior vena cava syndrome

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Introduction

Superior vena cava syndrome (SVCS) is a medical emergency, which in 80% of cases is caused by malignant mediastinal tumours. However, non-malignant causes lead to 20% of cases of SVCS.[1] We present an interesting case of SVCS after receiving ChAdOx1 CoV-19 vaccine (AstraZeneca).

Materials and methods

A 52-year-old man presented with progressively worsening swelling of the face, neck, chest and arms, pleuritic chest pain, abdominal pain, and breathlessness for 7 days. He had type 2 diabetes mellitus and was on gliclazide, metformin and sitagliptin. He received ChAdOx1 CoV-19 vaccine 5 weeks prior to his presentation. Physical examination showed a classical picture of superior vena cava occlusion with collateral vessels on the chest and reduced breath sound on the right base. Routine bloods showed raised inflammatory markers with white cell count 16.4 $\times$ 10^9/L, neutrophil count 13.53 $\times$ 10^9/L and C-reactive protein 44 mg/L, with a platelet count 338 $\times$ 10^9/L and D-dimer 3.25 µg/mL fibrinogen equivalent units, fibrinogen 4.1 g/L. Computed tomography of the neck, thorax, abdomen and pelvis including pulmonary angiography were done with no evidence of malignancy or vessels compression (Fig 1). Compression duplex ultrasound of upper limbs and neck revealed bilateral inferior jugular vein thrombosis, bilateral subclavian vein thrombosis extending into proximal axillary veins and superior vena cava thrombosis, right sided pleural effusion, no pulmonary embolism. Prothrombin time, activated partial thromboplastin time, renal and liver function tests, and C3, C4 and IgG4 levels were unremarkable. Vasculitic screen, connective tissue disease screen, anticardiolipin antibody, anti-beta2-glycoprotein antibody, lupus anticoagulant, PNH and JAK2 mutation were all negative. Anti-platelet factor 4 antibodies (anti-PF4) was positive. He was managed with low-molecular-weight heparin with good response and was switched to apixaban with a follow-up appointment in the deep venous thrombosis clinic.

Results and discussion

The patient's platelet count was persistently normal with a moderately raised D-dimer. Generally, patients with VITT present within 5–30 days post vaccination and thrombocytopenia (platelet count $<$150,000 $\times$ 10^9 L), D-dimer $>$4 µg/mL FEU, positive anti-PF4 antibodies on ELISA and presence of thrombosis.[2] In VITT IgG antibodies that recognise PF4 bound to platelets leading to widespread platelet activation.[3] Besides thrombosis and positive anti-PF4, our patient did not have any other features of VITT (although about 5% of patients with VITT typically have normal platelet count).[4] Moreover, in VITT, the

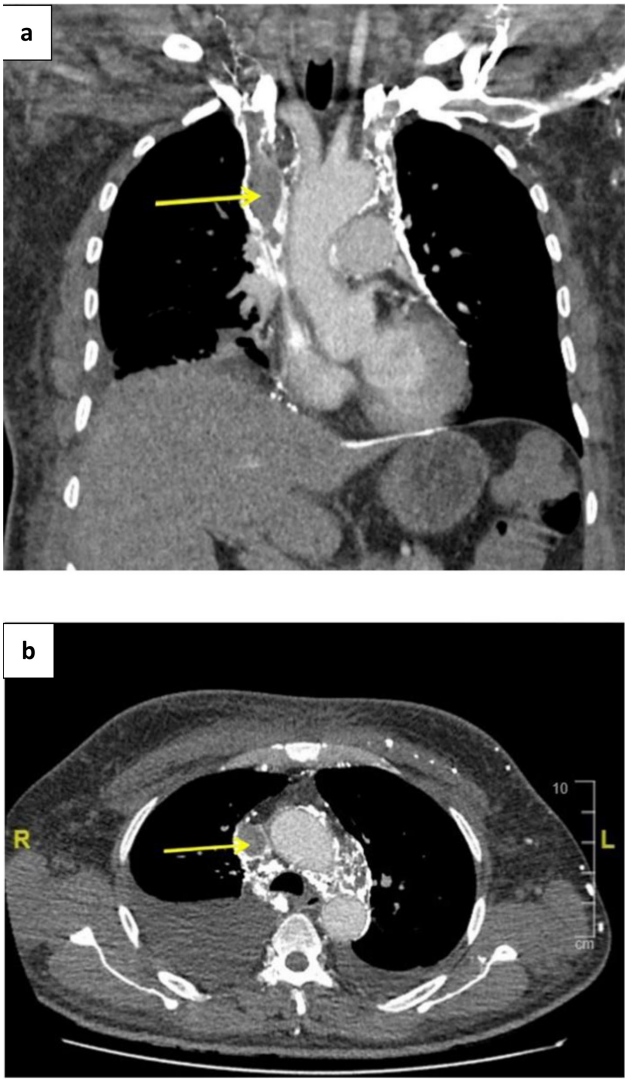

Fig 1. Computed tomography of the thorax. a) Coronal view: yellow arrow showing the thrombus in the superior vena cava. b) Axial view: yellow arrow showing the thrombus in the superior vena cava.

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cerebral vein, deep veins of the legs, pulmonary arteries and portal circulation are commonly affected by thrombosis,[5] which did not occur in our patient; rather, jugular, subclavian, axillary vein and superior vena cava were involved, causing SVCS.

Conclusion

Although exceedingly rare, VITT can be life-threatening, with a mortality rate of 22%,[2] thus highlighting the importance of not missing a diagnosis. This case report highlights the fact that all not cases of VITT have all the diagnostic features. Moreover, we think this is the first case report of VITT where the patient presented with SVCS. Therefore, clinicians should be vigilant when patient presents with thrombosis in atypical site and has had a history of recent COVID-19 vaccination to avoid missing this life-threatening complication. ■

References

1 Parish JM, Marschke RF, Dines DE. Etiologic considerations in superior vena cava syndrome. *Mayo Clin Proc* 1981;56:407–13.
2 Pavord S, Scully M, Hunt BJ *et al.* Clinical features of vaccine-induced immune thrombocytopenia and thrombosis. *N Engl J Med* 2021;385:1680.
3 Greinacher A, Selleng K, Palankar R *et al.* Insights in ChAdOx1 nCoV-19 vaccine-induced immune thrombotic thrombocytopenia. *Blood* 2021;138:2256.
4 Gabarin N, Sarah Patterson S, Pai M *et al.* Venous thromboembolism and mild thrombocytopenia after ChAdOx1 nCoV-19 vaccination. *Thromb Haemost* 2021;121:1677.
5 Greinacher A, Thiele T, Warkentin TE *et al.* Thrombotic thrombocytopenia after ChAdOx1 nCov-19 vaccination. *N Engl J Med* 2021;384:2092.