Pemberton's sign in SVC syndrome from metastatic renal cell carcinoma

Neil Keshvani, Christina Yek, David H Johnson

Department of Internal Medicine, University of Texas Southwestern, Dallas, Texas, USA

Correspondence to

Dr Neil Keshvani,
neil.keshvani@gmail.com

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DESCRIPTION

A 52-year-old man with no medical history presented to the emergency department after a syncopal episode. The patient reported reaching over his head to lift an object at work when he felt transiently light-headed, lost consciousness and fell to the floor. He denied any preceding palpitations or chest pain. He reported intermittent light-headedness and nausea for the past week, facial and neck swelling for several weeks, and an unintentional 20-pound weight loss over the past 3 months.

On physical exam, his temperature was 37.0° C, heart rate was 105/min, respiratory rate was 14/min and blood pressure was 137/84 mm Hg. He had mild neck and facial erythema and oedema, which became significantly more pronounced on elevation of the arms over the head (figure 1). A CT angiogram of the chest (figure 2) revealed a 6.9×4.7×5.8 cm mass in the mediastinum causing mass effect on the anterior aspect of the right main pulmonary artery and occluding the superior vena cava (SVC). The CT chest also revealed a partially imaged soft tissue mass arising from the upper pole of the right kidney, and dedicated CT abdomen and pelvis revealed a large 10.8 cm right renal mass. A biopsy of the chest mass subsequently demonstrated pathology suggestive of renal cell carcinoma. The patient was diagnosed with metastatic renal cell carcinoma causing SVC syndrome.

SVC syndrome is the obstruction of blood flow through the SVC. In the past, infectious aetiologies including syphilis and tuberculosis were a prevalent cause of SVC syndrome.¹ However, malignancy now far exceeds infection as the most common cause of SVC syndrome; of causative malignancies, primary lung cancer is the most likely, followed by lymphoma. Other non-malignant causes include thrombus secondary to

Figure 2 CT angiogram of the chest revealing a mediastinal mass causing compression of the superior vena cava.

Learning points

- ▶ Pemberton's sign involves bilateral arm elevation, and a positive sign results in facial engorgement. The exam manoeuvre is useful for detecting superior vena cava (SVC) obstruction.
- ▶ Renal cell carcinoma is a rare cause of SVC syndrome from tumour metastasis compression of the vein.

vascular catheterisation, goitre or mediastinal fibrosis.¹ The patient suffered from metastatic renal cell carcinoma, which is a rare and atypical cause of SVC syndrome.

Diagnosis of SVC syndrome is based primarily on imaging, but physical exam can help identify venous obstruction. Pemberton's sign (figure 1) was first reported by Dr Hugh Pemberton in 1946 and is characterised by facial plethora and venous engorgement following bilateral arm elevation.² This phenomenon is attributed to clavicular movement causing a 'nutcracker' effect that compresses major venous structures and was traditionally associated with large substernal goitres.³ This physical exam finding is a quick manoeuvre that can be beneficial in diagnosing SVC syndrome from all aetiologies.

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Figure 1 Pemberton's sign with elevation of the bilateral arms.

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REFERENCES

- 1 Wilson LD, Detterbeck FC, Yahalom J. Superior Vena Cava Syndrome with Malignant Causes. *N Engl J Med Overseas Ed* 2007;356:1862–9.
- 2 Pemberton HS. SIGN OF SUBMERGED GOITRE. *The Lancet* 1946;248:509.
- 3 De Filippis EA, Sabet A, Sun MR, *et al.* Pemberton's sign: explained nearly 70 years later. *J Clin Endocrinol Metab* 2014;99:1949–54.

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