Pemberton's Sign Revisited after 75 Years

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INTRODUCTION

Hugh Spear Pemberton was an English physician qualified from the University of Liverpool and started working at the David Lewis Northern Hospital in Liverpool. During the First World War, he served in the Royal Army Medical Corps, and he returned to the Northern Hospital only after the war. He then became a member of the Royal College of Physicians in 1921 and was appointed as consultant in 1924. He published articles on diabetes, thyrotoxicosis, and peripheral vascular disease. He had a sudden death 10 years after describing this eponymous sign.¹

PEMBERTON'S MANOEUVRE

Pemberton's maneuver reported first in 1946 is a useful clinical test for latent superior vena cava syndrome caused by a retrosternal mass.² With no clear consensus regarding the definition of retrosternal goiters, most authors follow the definition of DeSouza and Smith (1983) with more than 50% of the gland lying below the thoracic inlet. More recent papers³ including Hedayati and McHenry⁴ describe any goiter that extends below the thoracic inlet as a retrosternal goiter with the definition having an impact on preoperative planning and intraoperative need for sternotomy. Clinically oriented diagnosis needs imaging studies for confirmation. Computer tomography study is the gold standard for preoperative surgical planning.

Textbooks^{5–7} and case reports^{8–10} explain Pemberton's sign as a result of arm elevation causing "plugging" and obstructing the thyroid gland and its surrounding structure against the bony thoracic inlet or the so-called thyroid "cork effect." Thyroidal extension below the upper border of manubrium/thoracic inlet is defined as retrosternal goiter which connotes the cork has already crossed the inlet. Theory of cork effect with thyroid gland descent in to thoracic inlet (considered to be narrowest part in to thorax or mediastinum) during inspiration impeding the venous return in retrosternal goiters seems questionable.

De Filippis et al.¹¹ demonstrated that there was no craniocaudal movement of the goiter relative to the thoracic inlet when magnetic resonance angiography of the neck was performed while the patient's arms were elevated and at his sides, though the lateral aspect of the clavicle moved medially and inferiorly, obstructing the right external jugular vein and subclavian vein confluence compared to the movement of a "nutcracker" compressing major venous structures within a narrowed thoracic inlet against a relatively fixed and enlarged thyroid.

The above study¹¹ reiterates that when eliciting Pemberton's sign, facial plethora and venous engorgement were due to the clavicles moving and compressing venous vasculature against the

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enlarged thyroid and not to a "cork effect," though physiological compression of subclavian vein on arm abduction is identified in normal healthy volunteers.^{12,13}

Several studies that used CT and MR imaging in healthy volunteers to elucidate the exact mechanism of compression of neurovascular structures at thoracic outlet/inlet have demonstrated that upper-limb elevation does not induce any obvious change in the inter scalene triangle though it can cause physiological compression of subclavian vein upon arm elevation.¹² In a study¹³ for assessing detailed anatomy of thoracic outlet, universal venous compression is noted on helical CT at scalene clavicular and costoclavicular spaces in normal subjects upon abduction of upper extremity. Impending venous obstruction with possible compromise of collaterals in costoclavicular space, retro pectoral space, and inter scalene triangle along with increased venous pressure and drainage upon arm elevation in a retrosternal goiter may manifest with cyanosis and facial plethora.

Mechanism

We have used two figures to describe the mechanism of events at the thoracic inlet, leading to symptoms, such as dyspnea, flushing, and cyanosis, in an analogy to an express highway under an arched type of bridge. The entry climb of the bridge and exit descent can be compared to clavicles in the neck. The deck of the arched bridge represents the manubrium sterni. Below that is the neck chest line which divides the neck and mediastinum. The express highway can be divided into a prevertebral space, a carotid space, a visceral space, and a danger space. We propose that the retrosternal extension (RSE) of the goiter usually occupies the visceral space and may narrow the danger space when arms are elevated causing impedance to the venous return and thereby the symptoms (Figs 1 and 2).

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Fig. 1: Express way autologous representation of neck and mediastinum



Fig. 2: RSE compromising the danger space

CONCLUSION

Even though more than 75 years have elapsed after the first description of this sign, the scientific basis behind the manifestation is not clear and may also include other factors, such as neck type (short neck), muscularity of the individual, and also other factors. The sign may cause respiratory distress and may not be elicited when other features or imaging evidence is present.¹⁴

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