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The chest X ray in pulmonary embolism: Westermark sign, Hampton´s Hump and Palla´s sign. What´s the difference?

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ABSTRACT

Pulmonary embolism (PE), with the incidence of about 60 per 100 000 annually, can be a life-threatening disease if it is not treated promptly. It has been estimated that some 10% of PE patients die within the first hour of the event. Untreated PE has a mortality of about 30%. PE is a condition that is treatable if suspected and diagnosed early. The chest radiograph is still the first investigation that is ordered in patients presenting with cardiorespiratory symptoms or symptoms suggestive of PE. The CXR is also helpful in identifying or excluding other conditions or diagnoses. Thus, knowing and understanding some of the more specific CXR signs can be useful. We suggest that physicians to be aware of and utilize CXR findings such as Palla's sign, Westermark sign and Hamptons hump to help with the diagnosis of PE and to exclude other conditions that can mimic venous thrombo-embolism. Even if these signs are not common, their presence, even in an unsuspected patient without a high pretest probability of PE, should prompt further investigations such as a D-dimer test, lung scintigraphy or computed tomography pulmonary angiography as required.

1. Introduction

Pulmonary embolism (PE), with the incidence of about 60 per 100 000 annually, can be a life-threatening disease if it is not treated promptly. It has been estimated that some 10% of PE patients die within the first hour of the event. Untreated PE has a mortality of about 30%[1-3].

PE is caused by an embolic obstruction of the pulmonary arteries which can impair blood flow to the lung leading to a ventilation-perfusion (V/Q) mismatch. This can then result in a spectrum of cardiorespiratory complications from hypoxemia to cardiac arrest depending on the size and the chronicity of the emboli. The range of symptoms of patients with PE can range from pleuritic and non pleuritic chest pain, dyspnoea, cough, haemoptysis, syncope, to a collapsed state. The signs also have wide variations and can include tachypnoea, tachycardia, hypoxia, cyanosis, fever and crepitations in the lungs[3-6].

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Bearing this in mind, even with better diagnostic equipment at hand, PE remains as one of the most challenging diagnosis for front line physicians to make. About 90% of the embolus originate from deep vein thrombosis of the proximal lower limbs and pelvis. From another perspective, about 50% of deep vein thrombosis in the legs embolize to the lung. The risk factors of venous thromboembolism can be attributed to any condition which result in an abnormality of any component of the Virchow's triad: [6-10]

(1)Stasis in blood flow, such as immobilisation post-surgery or during long haul flights

(2)Changes in the endothelium, from direct wall injury or other causes

(3)Hypercoagulable states which may occur with malignancies, oral contraceptives or thrombophilia.

Investigations would then be conducted based on the clinical probability of PE. The National Institute for Health and Clinical Excellence guidelines advocate the use of a diagnostic algorithm that incorporates the pretest probability two-level Wells PE score and D-dimer assay results (in appropriate patients) to determine the use of subsequent diagnostic tests. A low probability of less than 4 points for 2 tier Wells' Criteria means that performing a plasma D-dimer is sufficient to rule out any possibility of PE. This is due to the high sensitivity (up to 94%) but low specificity (up to 45%) of the test. Therefore, having a negative test reliably excludes PE[1,8,11,12].

In the past or in less developed countries without easy access to CT machines, lung scintigraphy is performed to obtain the diagnosis of PE by observing for any V/Q mismatches[1,3,4]. Revised PiOPED criteria reported a sensitivity of 41% and specificity of 97%. However, there is a large percentage of scans in suspected patients that actually fall in the category of intermediate probability of PE and hence further work up will be needed[13]. At present, such V/Q imaging is only indicated in PE with patients that has contraindications to CT imaging such as renal failure and contrast allergy[6,10,13]. With the advance of technology, the current gold standard of clinching the diagnosis of PE is computed tomography pulmonary angiography. The famous PIOPED 2 study and the British Thoracic Society both advocate that it should be used as the first line of investigation in all patients with a high clinical probability of PE. The sensitivity of it is 83% and has a specificity of up to 100%. It can also be used to rule out other differentials like aortic dissections[11-15].

Indeed having a good history, physical examination would pave the way for pre-test probability (using Wells' or PERC) and subsequent investigations to be done. However, certain atypical presentations of PE might confer a low clinical suspicion and cause investigations not be conducted, but in a patient who presents with shortness of breath, a chest X-ray (CXR) is usually a routine investigation in emergency departments globally to ascertain the cause. There are characteristic features of PE that can sometimes be found on CXR that can help with the diagnosis. Although the usefulness of such features are constantly under debate[3,8,10,14-17].

Important features such as Hampton's hump, Palla's sign and Westermark sign can be easily confused. In this paper, we aim to provide a clearer picture of each sign and further insights into their usefulness especially since most (if not all) dyspnoeic patients in the emergency department will get a CXR done[16-19].

2. Chest radiography in suspected PE

CXR is widely used in all emergency departments around the world as the first line investigation for all suspected cardiopulmonary conditions. It is readily available and is used as a diagnostic modality for certain conditions such as acute pulmonary edema, pneumonia and pneumothorax. The CXR serves as an initial risk stratification tool as well. It thus pays to be able to read it well. In some institutions, radiologist consult is available 24 h for clinicians to discuss even the most subtle signs. In the usual circumstances, the CXR is done as a posterior-anterior view but in the ICU and more acute settings, mobile CXRs done with the suboptimal PA view, may make it even more challenging to pick up the subtle radiological signs[10,14,16,17-21].

There has been much discussion on the reliability of CXR features in PE. In a review of 1 063 patients with suspected PE, only 12% of those proven to have PE were found to have normal chest radiograph findings[18]. The international cooperative study of the PE registry reported that only 24% of 2 452 patients with acute PE had normal chest radiograph findings[19]. In another study done with 50 patients, only 18% had normal chest radiograph findings[20]. Based on such numbers, it is estimated that about 80% of patients with acute PE had an abnormal CXR. The most common abnormality noted from 4 studies was cardiomegaly[19,20]. The other signs frequently noted were pulmonary infitrates, atelectasis, pleural effusion, pulmonary congestion, elevated hemidiapgram, Palla's sign, Westermark sign and Hampton's hump. Although cardiomegaly is the most frequent finding in up to 38%, it cannot be used to establish the diagnosis due to the long list of possible differential diagnoses including heart failure, pericardial effusion, hypertrophic heart disease and severe valvular lesions. Such conditions are also prevalent worldwide and even more so in the future, with the advancements of medical treatment and resultant increase in life expectancy of patients with these conditions[22-24]. Other CXR findings of PE are also non-specific as certain lung pathologies can also have similar features. However, there are certain signs that have a higher specificity and they are the Palla's sign, Westermark sign and Hamptons Hump[25-28].

3. Westermark's sign

Westermark's sign refers to a focal area of enhanced or increased translucency due to oligaemia, which occurs due to impaired vascularisation of the lung due to primary mechanical obstruction or reflex vasoconstriction. The sign is formed by dilatation of the pulmonary arteries proximal to the site of emboli followed by a

sharp and demarcated collapse of the distal vasculature[28-35].

The Westermark's sign is rare and was only found in 8%-14% of confirmed pulmonary embolism cases in the PIOPED study[28-31]. However, it is highly specific and should raise one's suspicion of pulmonary embolism if present. A study by Risti L found out that in patients with chronic hypoxemic and secondary erythrocytosis, the presence of Westermark's sign on radiological imaging conferred a 2.286 times higher probability of having pulmonary embolism than other similar patients without the sign[36].

Accuracy in the interpretation of the sign can be enhanced by comparing the current chest radiograph with the patient's previous chest x-ray films. It is also difficult to visualise the Westermark's sign when the chest x-ray is performed in a supine position[18,28,32,34].

4. Palla's sign

Palla's sign refers to an enlargement of the right descending pulmonary artery proximal to a cut off of the pulmonary artery due to acute pulmonary embolism. This sign was first described in 1983 by Palla A, whereby the typically "sausage" appearance of the descending pulmonary artery was seen in 25% of the patients with confirmed pulmonary embolism and not present in patients without pulmonary embolism[29,30].

The Palla's sign is ascertained by measuring the diameter of the right descending pulmonary artery at the superior venous angle, then distally at 10 mm, 20 mm and 30 mm from the superior venous angle. Palla's sign is established when the diameter of the right descending pulmonary artery is more than 16 mm at the superior venous angle[29-34].

The Palla's sign has a low sensitivity and unknown specificity. Although the sign is rare, it is still valuable in aiding the diagnosis of pulmonary embolism when seen with other signs like the Westermark's sign, the Hampton hump, and the Fleischner sign (dilated pulmonary artery)[30,32,34,37].

The combination of Palla's and Westermark's sign can suggest an occlusion of a lobar or segmental pulmonary artery by an emboli or widespread occlusion in multiple small arteries[29,31,32].

5. Hampton's hump

Hampton's hump is seen on the chest radiograph as a wedge-shaped opacity with a rounded convex apex directed towards the hilum. The Hampton's hump occurs within two days of a pulmonary infarction, whereby subsequent alveolar necrosis and hemorrhage into an incomplete infarct accounts for the opacity. After a few months, the pulmonary infarct resolves and a residual scar remains[6,16,35].

Even though the Hampton's hump has a high specificity of 82%, it has a low sensitivity of 22% which limits its usefulness in the diagnosis of pulmonary embolism[33-35]. The low sensitivity of the sign can be explained by the dual blood supply of the lungs, which is present in majority of the people. With collateral vascular supply from both the pulmonary and bronchial arteries, the bronchial arteries protect against a pulmonary infarction in the event of a pulmonary embolism[33,35-38].

The Hampton's hump is seen more commonly in patients with certain co-morbidities affecting the cardiopulmonary system like chronic obstructive pulmonary disease, left heart failure and venous pulmonary hypertension[30,33,36]. The Hampton's hump is also more commonly seen in the lower lobes and often associated with pleural effusion[26,27,33]. However, the Hampton's hump can sometimes be misdiagnosed as pneumonia with an alveolar consolidation. Hence, importance should be placed in the ability to accurately recognise the sign on chest radiographs[33,35].

6. Recommendations

PE is a condition that is treatable if suspected and diagnosed early. The chest radiograph is still the first investigation that is ordered in patients presenting with cardiorespiratory symptoms or symptoms suggestive of PE. The CXR is also helpful in identifying or excluding other conditions or diagnoses. Thus, knowing and understanding some of the more specific CXR signs can be useful. We suggest that physicians to be aware of and utilize CXR findings such as Palla's sign, Westermark sign and Hamptons hump to help with the diagnosis of PE and to exclude other conditions that can mimic venous thrombo-embolism. Even if these signs are not common, their presence, even in an unsuspected patient without a high pretest probability of PE, should prompt further investigations such as a D-dimer test, lung scintigraphy or computed tomography pulmonary angiography as required.

Conflict of interest statement

The authors report no conflict of interest.

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