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Pneumomediastinum and pneumorrhachis: a lot of air about nothing?

Case report

Statement of interest
None declared.

A never-smoking 22-year-old male with no past medical history presented to the emergency department with sudden onset of sharp central chest pain and right-sided neck discomfort. There was no history of trauma or bronchitis. There was no history of air travel, allergen exposure or illicit drug use. He had consumed alcohol the night before presentation but had not vomited.

Clinically, he had right-sided subcutaneous emphysema but no signs of pneumothorax. His pulse was 82 beats per min, blood pressure was 134/76 mmHg and oxygen saturation was 98% on air. His blood tests revealed no abnormality. His blood alcohol level was not measured.



Figure 1
Chest radiography.

Task 1

Interpret the chest radiograph (fig. 1)

HERMES syllabus link:
module B.13.3

Answer 1

The chest radiograph (fig. 1) shows right-sided subcutaneous emphysema in the neck and a pneumomediastinum (PM) with air tracking in the muscle planes of the neck but no pneumothorax.

At presentation, a diagnosis of oesophageal rupture was considered but an upper gastrointestinal endoscopy was normal. Contrast-enhanced helical computed tomography (CT) of the thorax and upper abdomen was performed (figs 2–5).

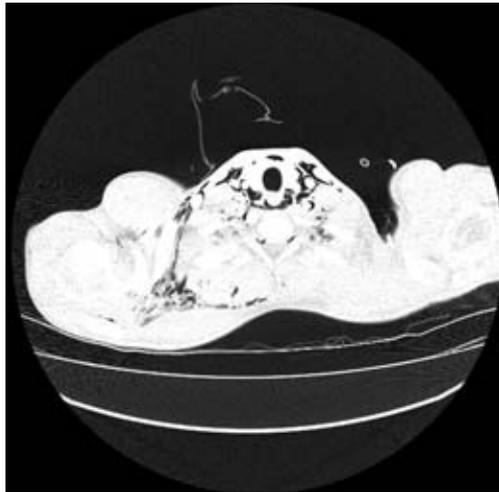


Figure 2
Contrast-enhanced helical CT.



Figure 3
Contrast-enhanced helical CT.



Figure 4
Contrast-enhanced helical CT.



Figure 5
Contrast-enhanced helical CT.

Task 2

Interpret the CT images (figs 2–5)

Answer 2

The CT images show extensive subcutaneous emphysema on the right, extensive PM outlining the vessels with air, and air in the spinal canal, known as pneumorrhachis (PR). There is no pneumothorax or oesophageal rupture.

His symptoms settled with conservative management and he remained cardiovascularly stable, such that he was discharged. He remained very well on follow-up 4 days later, he had no symptoms and his surgical emphysema had reduced considerably. On final follow-up after 3 months, his chest radiograph had completely normalised.

This is the first case of PR in the context of PM that we have encountered. It is a rare clinical entity that should be recognised by all physicians and, hence, we discuss the aetiology and management of both.

Discussion

Free air in the spinal cord is an uncommon phenomenon that was first reported in 1977 [1]. Various names have been attributed to this phenomenon (intraspinal pneumocele, spinal epidural and subarachnoid pneumatosis, spinal and epidural emphysema, aerorachia, pneumosaccus, and air myelogram) until, in 1987, the term PR was universally applied [2].

PR is classified descriptively into an internal, intradural or external, extradural pattern. Internal traumatic PR is frequently associated with major trauma and believed to be a marker of severe injury [3]. Penetrating spinal injuries are usually a direct source of air entry, but other causes of PR can be explained by entrapment of air into the paraspinal soft tissues and into the epidural space *via* the neural foramina along the vascular and nerve root bundles. This could occur in conditions producing exceptionally high intrathoracic pressures (*e.g.* coughing, sneezing and vomiting) [4–7] as well as either a complication of surgical or anaesthetic procedures or a complication of malignancy and its therapy [2].

Our patient had a PM associated with PR, along with subcutaneous or surgical emphysema. The association between PM and surgical emphysema as a result of barotrauma is well known. It can occur spontaneously or in association with coughing, vomiting or sports. High bronchoalveolar pressures can

lead to air leakage into the mediastinum through the bronchovascular layers. The air dissects the path of least resistance onto the fascial planes of the neck, a process that is known as the Macklin effect [8]. Less understood is the association of PM with PR. There are no fascial barriers between the posterior mediastinum and the retropharyngeal and the epidural spaces, which allows free diffusion of air into the epidural space through the posterior surface due to the lower resistance of the connective tissue as opposed to the anterior vascular bundle.

There are no epidemiological data but a review of the literature [9] describes 71 case reports or series, each encompassing no more than three cases reporting a total of 86 cases of PR. A more recent review by BELLOTI *et al.* [10] found 42 reports for 47 cases of spontaneous PM complicated by PR that were published between 1989 and 2009. The median age of occurrence was 18 yrs and there was a male predominance in the epidemiology. The diagnosis was always made on CT scanning. In five of the cases, PM and PR were associated with a pneumothorax. 14 patients had an underlying lung disease, most notably asthma. Vomiting, coughing, air travel, illegal drug use and hyperventilation were associated with 24 of the cases. 45 of the patients were treated conservatively and contrast-swallow did not demonstrate oesophageal rupture in any of the patients with vomiting.

As such, invariably, PR is found in combination with associated air distribution in other compartments and cavities of the body: particularly in conjunction with pneumocephalus, pneumothorax, PM, pneumopericardium or subcutaneous emphysema. It is usually found incidentally on CT or magnetic resonance imaging [11–14]. The differential diagnosis of intraspinal air is wide and can be due to degeneration, inflammation, malignancy and infection [15].

PR tends to be largely asymptomatic, as the air is eventually reabsorbed into the circulation without any risk of re-accumulating. Rarely, PR can cause symptoms; there was a progressive motor deficit of the lower limbs due to introduction of intraspinal air in a patient who requires repeated lumbar punctures [16]. The patient had to be operated on for closure of the dural defect. Because of the rarity of PR, no international guidelines exist for its management and each case must be considered individually within

a multidisciplinary setting. There is only one case report of a patient without underlying lung disease who needed a laminectomy for progressive neurological signs and symptoms for a massive PM and PR [17].

As mentioned, our patient settled with conservative management and repeat imaging showed complete resolution of PM and surgical emphysema and he was discharged from follow-up.

Key learning points

- Young male patients without underlying lung disease are most at risk of developing spontaneous PM and PR.
- With the increasing use of CT scanning, the reported incidence of PR may rise.
- PM associated with PR has a benign course in the vast majority of patients and patients can be expected to recover fully.
- Neurological signs in these patients should be acknowledged seriously but no accepted guidelines for their management exist.

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