PNEUMOMEDIASTINUM AND PNEUMORETROPERITONEUM ASSOCIATED WITH PERFORATED ESOPHAGEAL CANCER

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ABSTRACT

Pneumomediastinum (PM) and Pneumoretroperitoneum (PRP) may rarely result from esophageal rupture which is a potentially life threatening condition resulting most frequently from instrumentation. A middle aged male presented with dysphagia and weight loss. Oesophagoscopy revealed a tight complex stricture, that was dilated with CRE balloon to facilitate further assessment of stricture anatomic details, following which the patient developed PM, PRP and subcutaneous emphysema (SE). The endoscopic biopsy from the proximal stricture area was reported as epithelial malignancy. A diagnosis of esophageal malignant stricture with iatrogenic perforation was rendered and total oesophagectomy was performed which revealed a circumferential transmurally infiltrating keratinizing squamous cell carcinoma (Grade 2) involving the distal third of the esophagus. A cautious approach should be employed while evaluating patients with narrow and complex strictures. In these cases a tissue diagnosis is desirable before institution of esophageal procedures. Critical post procedural monitoring for PM, PRP and SE is imperative for prompt diagnosis of esophageal perforation as the rate of mortality and complications depend on the interval between initiation of the procedure and perforation.

Key words: Esophageal perforation, pneumomediastinum, pneumoretroperitoneum, subcutaneous emphysema

INTRODUCTION

Pneumomediastinum (PM) connotes the presence of free air or gas in the mediastinum that almost invariably originates from the alveolar space or conducting airway. Esophageal rupture is an uncommon cause of PM. It is a rare potentially lethal event resulting most frequently from instrumentation of tight, fibrous or complex esophageal strictures, thereby making thorough patient evaluation before instrumentation, a necessity.
We present a case of combined PM, PRP and SE secondary to iatrogenic perforation of an esophageal stricture that was subsequently diagnosed as locally advanced squamous cell carcinoma, and emphasize the importance of employing a cautious approach in evaluation of patients with tight or complex stricture.

CASE REPORT

A 55 year old male presented with progressive dysphagia for solid food and loss of appetite for the past 6 months associated with mild to moderate nonradiating epigastric pain and significant weight loss. On examination he was poorly built and nourished. Per abdominal examination revealed mild epigastric tenderness and no organomegaly or mass was discerned. The full hemogram, renal and liver function tests, chest x-ray and abdomino-pelvic ultrasound were normal. Esophageal endoscopy revealed a tight “complex” stricture at 35cm that did not allow further advancement of the scope. A superficial biopsy from the proximal accessible portion of the stricture was performed. In order to fully assess the anatomic details and facilitate further biopsies, dilation with 15mm CRE balloon was attempted.

Figure 1 A. Chest radiograph (PA) showing surgical emphysema in the neck (red arrows) and axilla (yellow arrows).
Figure 1 B. Abdominal radiograph (erect) showing pneumoretroperitoneum (red arrows).

Figure 2 A. CT scan showing cervical emphysema (red arrows).
Figure 2 B. CT scan (lung window) showing pneumomediastinum (red arrow) and minimal right pneumothorax (yellow arrow).

Figure 2 C. CT scan (mediastinal window) showing oesophageal thickening (yellow arrow) minimal right pleural effusion (red arrow).
Figure 2 D. CT scan showing pneumoretroperitoneum (red arrows).

Figure 2 E. CT scan showing leakage of contrast (red arrow) with pneumoretroperitoneum (yellow arrow).
Following the dilation procedure the patient experienced retrosternal pain that progressively increased in severity. Associated swelling over neck and upper part of the left chest, that was crepitant on palpation, developed over a period of 2 hours. The heart and lung sounds were notably decreased and mediastinal crunch suggestive of PM was present on auscultation. Vitals were stable and arterial blood gas and basic metabolic parameters were normal. Complete blood count revealed neutrophilic leukocytosis (12,400 leukocytes/mm$^3$ with 80% neutrophils). Chest x-ray revealed SE in the neck, axilla and PRP (Figures 1A and 1B). Contrast enhanced computed tomography revealed diffuse irregular circumferential thickening of the lower third of the thoracic esophagus with moderate enhancement, diffuse extensive surgical emphysema involving the neck base, mediastinum and retroperitoneum, minimal right pneumothorax and pleural effusion and circumferential SE in bilateral chest (Figures 2A, 2B, 2C and 2D). There was extravasation of the oral contrast into the perigastresophageal region and retroperitoneum which was suggestive of esophageal perforation (Figure 2E). The patient was put on broad spectrum intravenous antibiotic therapy and total parenteral nutrition. Following the biopsy report of esophageal squamous cell carcinoma (SC Ca) a total oesophagectomy with gastric pull up and feeding jejunostomy was performed.
Macroscopy of the resected specimen revealed a grey white, friable, ill circumscribed ulceronecrotic growth m/s 5.5x 2.2cm circumferentially involving the distal esophagus, infiltrating the subjacent esophageal wall to a depth of 1.4cm with adventitial extension (Figure 3). A fistulous tract, bordered by inflammatory exudate, was situated on the posterior aspect of the tumor. Microscopy revealed a transmurally infiltrating moderately differentiated (Grade 2) SC Ca with local lymph node involvement, pT3 pN1 pM0, Stage 3 (Figures 4A and 4B).

Figure 4 A. Microphotograph showing oesophageal squamous cell carcinoma with nests and islands of malignant squamous cells (T) and intervening desmoplastic stroma (D). [H & E, x100]
DISCUSSION

PM results from diverse causes that may be intra thoracic (alveolar rupture, blunt chest trauma, straining against a closed glottis, narrowed/ plugged air way) or extra thoracic (sinus fracture, iatrogenic manipulation in dental extraction, hollow viscus perforation)\(^2\). The most common cause is alveolar rupture following sudden rise in intrapulmonary pressure with subsequent Macklin effect\(^1\). The air present in the mediastinum may dissect along periaortic and periesophageal fascial planes, sternocostal diaphragmatic attachment, retropharyngeal space, vascular sheaths within the neck and submandibular space resulting in PRP and SE\(^1\).

A relatively less common, albeit life threatening cause of PM is esophageal perforation (OP)\(^3\). The most frequent cause of the latter is iatrogenic (instrumentation) (43-55%) other causes being foreign body ingestion (7-14%), spontaneous (15%), trauma (10%), operative injuries (8%) and tumor (4%)\(^3\)-\(^4\). The underlying diseases in patients with OP were benign stricture (peptic, radiation, post surgery, corrosive) (22%), anatomic lesion (diverticula, webs, rings), tumor (10%) and motility disorders (achalasia) (5%) and 40% had no underlying esophageal pathology in one study\(^4\).
Esophageal dilation is a relatively safe procedure performed to relieve benign or malignant dysphagia. The main complications of the procedure include perforation, aspiration and bleeding. The rate of perforation following balloon dilation depends on the endoscopist’s experience, aetiology and complexity of stricture, type of dilator used and the technique employed. A regional audit in United Kingdom reported an overall perforation rate of 2.6% with mortality of 1%. Another study of 365 esophageal dilations revealed a perforation rate of 0%. Perforation was less common following dilation of benign strictures (1.1% with a mortality rate of 0.5%) than malignant stricture (6.4% with a mortality rate of 2.3%). Further SC Ca’s were more prone to perforation than adenocarcinomas (4.4% vs. 2%).

In the current case, the SC Ca was high stage at presentation and exhibited i) predominantly intramural growth with extensive central ulceration, ii) circumferential and transmural esophageal penetration, iii) florid desmoplastic response, iv) irregular infiltrative growth rather than expansile growth and v) extensive necrosis with acute inflammatory reaction, all of which presumably facilitate perforation when radial forces are generated by balloon dilation.

Balloon dilation is used to facilitate diagnostic endoscopy when a stricture prevents passage of the endoscope. However if a stricture is narrow and complex with clinical suspicion of malignancy, dilation is hazardous and a barium study may be used to provide anatomic details; if dilation is attempted it should be done under fluoroscopic guidance. Biopsy and dilation should not be performed during the same examination in such cases and a tissue diagnosis (biopsy or brush cytology) is desirable prior to dilation as it will influence overall management and estimation of perforation risk. Large calibre dilators (7.15mm) should be avoided and only modest dilation sufficient to permit biopsy or endoscopic ultrasound or facilitate stent insertion should be advocated preferably under wire guided or endoscopically controlled techniques.

OP should be promptly diagnosed as the rate of mortality and complications (mediastinitis) depend on the interval between initiation of treatment and perforation. The presence of neck, chest (acute retrosternal/pleuritic) or abdominal pain following endoscopy requires further investigation to rule out OP. Physical examination may reveal SE (crepitant on palpation), respiratory distress or Hamman sign (crunching sounds that are synchronous with the heart beat on auscultation) (pathognomonic for PM). Plain film may reveal features of PM (Naclero’s sign, continuous diaphragm sign, vertical and paratracheal streaks, ring around the artery sign or extra pleural air sign), pneumopericardium, pneumothorax or subdiaphragmatic air. CT may reveal extraluminal air and contrast oesophagograms are diagnostic.

CONCLUSION

Esophageal procedures should be adequately tailored to the underlying disease. Even though esophageal balloon dilation is a safe, minimally invasive and effective procedure for the management of strictures, it should be performed with utmost caution in patients with high probability of cancer, who usually have tight and complex strictures. In order to minimise complications, tight esophageal stenosis should be evaluated under fluoroscopic guidance.
LIST OF ABBREVIATIONS

Pneumomediastinum – PM; Pneumoretroperitoneum – PRP; Subcutaneous emphysema - SE; Esophageal perforation – OP; Squamous cell carcinoma - SC Ca

REFERENCES