Case report - Thoracic oncologic

Extended right pneumonectomy in an adult with a double aortic arch: a therapeutic dilemma

Stephan Christian Werth¹, Emma McLean⁰, Loïc Lang-Lazdunski*-*

¹Department of Thoracic Surgery, Guy’s and St Thomas Hospital, Great Maze Pond, London SE1 9RT, UK
²Department of Histopathology, Guy’s and St Thomas Hospital, Great Maze Pond, London SE1 9RT, UK

Received 28 June 2010; received in revised form 24 August 2010; accepted 27 August 2010

Abstract

We report the case of a 69-year-old man presenting with a primary right lung cancer and a complete double aortic arch. An extended right pneumonectomy was successfully performed and the patient remained well at the one-year follow-up. We discuss the surgical approach and the technical considerations imposed by this rare vascular abnormality.

© 2010 Published by European Association for Cardio-Thoracic Surgery. All rights reserved.

Keywords: Double aortic arch; Aortic arch anomaly; Lung carcinoma; Pneumonectomy

1. Introduction

Aortic arch anomalies are well-described in infants, causing stridor, respiratory distress or dysphagia in the first months of life [1]. Double aortic arch (DAA), has been abundantly described in childhood and its management is well-established [2]. Yet, DAA presenting in adulthood is rare and only a few cases have been reported [3]. Most publications report on symptomatic aortic arch anomalies. By contrast, asymptomatic vascular rings are often diagnosed incidentally during life or at postmortem. Hence, there is very little information on the outcome of patients with DAA after major lung resection. We present the case of a 69-year-old patient with an incidentally diagnosed DAA who underwent extended right pneumonectomy for lung carcinoma. To our knowledge, this is the first report of a pneumonectomy in an adult with DAA.

2. Case report

A 69-year-old male was referred to our thoracic unit for investigation of a lung mass. He had initially presented with symptoms of bowel obstruction and had had an emergency right hemi-colectomy for Dukes A adenocarcinoma three months previously. Chest radiography (X-ray) showed a right lung mass and enlargement of the superior mediastinum (Fig. 1b). Chest computed tomography (CT)-scan had revealed a tumor infiltrating the right hilum, together with a complete DAA of right-dominant type with left-sided descending aorta (Fig. 1a). The right subclavian artery arose from the right branch. The lung mass was biopsied under CT-guidance and showed a spindle cell carcinoma. Positron emission tomography (18-FDG-PET-CT) showed a right lung mass with a maximum standard uptake value (SUV_max) of 20 and a focus of increased tracer uptake was also noted in the distal esophagus. There was no abnormal uptake anywhere else. Esophagoscopy and biopsy demonstrated Barrett’s disease and primary adenocarcinoma of the distal esophagus. The patient underwent endobronchial ultrasound (EBUS), esophageal ultrasound (EUS) and cervical mediastinoscopy, all ruling out mediastinal lymph node involvement. Despite his excellent performance status the risk of combined pneumonectomy and esophagectomy was considered too high. He was scheduled for right pneumonectomy followed by radical chemoradiotherapy to treat the esophageal carcinoma. A right posterolateral thoracotomy was performed with en-bloc resection of the right lung, subcarinal lymph nodes and pericardium. Radical mediastinal lymphadenectomy was performed and the right main bronchial stump was covered with an intercostal muscle flap. Postoperative bronchoscopy showed no tracheobronchial compression. Postoperatively, the patient was discharged with a bronchopleural fistula which was successfully closed by the application of Bioglue® surgical adhesive (Cryolife Europa Ltd, Guilford, Survey, UK) through rigid bronchoscopy. The pleural cavity was debrided by video-thoracoscopy and a drain was left in for two weeks. The patient was finally discharged home on day 30.

Histopathology showed a purely myoepitheliomatous variant of epithelial–myoepithelial carcinoma. A tumor was found involving the visceral pleura and the hilar lymph nodes. Resection was complete. Final staging was pT3N1M0.

The patient received radical chemoradiotherapy for his early-stage esophageal carcinoma and was well and disease-free at one year. Serial chest X-rays and chest CT performed at seven months showed no tracheal compression (Fig. 1c). At last follow-up, there was no sign of bronchopleural fistula.

*Corresponding author. Tel.: +44 2071881038; fax: +44 2071881016. E-mail address: loic.lang-lazdunski@gstt.nhs.uk (L. Lang-Lazdunski).
© 2010 Published by European Association for Cardio-Thoracic Surgery
DAA usually presents in early infancy often accompanying cardiac defects [2]. In adulthood, DAA is mainly diagnosed during hospitalization for either respiratory symptoms or esophageal compression [3]. Our patient was completely asymptomatic. Preoperative chest CT demonstrated no significant narrowing of the trachea or esophagus. Thus, there was no indication for correction of the anomaly. Nakamura et al. reviewed the literature and reported six patients with right aortic arches undergoing lobectomies for primary lung cancer [4]. However, the level of mediastinal shift observed following lobectomy is much less than the one after right pneumonectomy. In addition, George et al. reported on a case of DAA who could not be extubated due to severe tracheal compression following a cardiac surgical procedure and required correction of the anomaly under cardiopulmonary bypass [5].

In our patient, we were particularly concerned about the risk of major mediastinal shift and subsequent compression of the trachea. Thus, excessive mediastinal shift following pneumonectomy potentially leads to rotation of the heart and great vessels inducing tracheal or bronchial compression (postpneumonectomy syndrome). This complication is more common following right pneumonectomy [6]. Interestingly, postpneumonectomy syndrome has been frequently observed in patients with a right-sided aortic arch following left pneumonectomy, by contrast only infrequently in patients with left-sided aortic arch [6]. In our patient, postoperative chest X-rays showed minimal mediastinal shift and bronchoscopy showed no tracheal or bronchial compression. One could speculate as to why the mediastinum did not shift significantly postoperatively. Of note, no ligamentum arteriosum could be found on the right when dissecting the proximal right pulmonary artery. In this case, we considered inserting an implant in the pleural cavity to prevent postpneumonectomy syndrome, but this proved unnecessary. Alternative options included dividing the minor left aortic arch and performing the right pneumonectomy through a median sternotomy. This approach seemed hazardous and priority was given to the best exposure for complete excision of the lung tumor.

The other remarkable finding in this case is that the patient was diagnosed with three different malignant tumors in a short time frame. The chest X-ray done prior to colectomy helped diagnose the lung mass and the PET-CT performed in the work-up of the lung carcinoma helped diagnose the esophageal carcinoma. The patient was very fit and the multidisciplinary meeting decision was to offer him curative treatment for his thoracic cancers. The lung investigations were delayed until the patient had recovered from his colonic resection. We did review the original chest X-ray which was indeed much less impressive than the one shown on Fig. 1, taken three months later, and showed a small tumor in the right upper lobe. Obviously, we had to wait until the patient had recovered from his colectomy to perform the right pneumonectomy.

References


