Case Report.

Metastatic Oesophageal Adenocarcinoma to an Intralobar Bronchopulmonary Sequestration: Histopathology Perspective.

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Abstract

Broncho-pulmonary sequestration is a rare anomaly which represents 0.15- 6.4% of all pulmonary malformations. (1, 2) It is defined as the presence of non-functioning lung tissue that is separate from the bronchial system and is supplied by an aberrant artery originating from the aorta or one of its branches (1, 2). Bronchopulmonary sequestration is divided into two types: Intralobar; which shares the same pleura of the normal lung, and extralobar, which is covered by separate pleura.

Pulmonary sequestrations can be asymptomatic but they may cause recurrent infections and respiratory symptoms such as productive cough, haemoptysis or chest pain. (3) Primary malignancy arising in pulmonary sequestrations is rare with very few reported cases; none of these described metastatic carcinoma.

This report describes a case of oesophageal adenocarcinoma, metastasising to an intralobar bronchopulmonary sequestration. To my knowledge, this is the first documented case of a tumour metastasising to pulmonary sequestration.

الخلاصة

الانحجاز الرئوي هو عبارة عن كتلة من الشعبيات اليوانية المنفصلة عن الجهاز التنفسي. قد تشارك هذه الكتلة الانحجازية مع الرئة الطبيعية بنفس الغشاء، أو تكون مغطاة بغشائها الخاص. في كلا الحالتين يتمتد الانحجاز الدم من الشريان الأورطي أو أحد فروعه بدلاً عن الشريان الرئوي مما يجعل الانحجاز منفصلاً عن الرئة تشريحياً ووظيفياً. في كثير من الأحيان لا تنشأ أعراض مرضية بسبب الانحجاز لكنه قد يشكل بؤرة للالتهابات مما يؤدي البلغم بالإضافة إلى المضاعفات التنفسية تشمل السعال والصدراوء نزيف رئوي. هناك عدد ضئيل من التقارير الطبية المشتركة التي توثق حدوث سرطان في الانحجاز الرئوي. كلما اصف اوراماً أولية ناشئة عن الانحجاز نفسه. هذا التقرير يصف حالة ورم ثانوي أصاب الانحجاز الرئوي حيث انتشر سرطان الرئة عند سيدة في الخمسين من العمر إلى الانحجاز الرئوي.

Case Report

A 50 year old, ex-smoker woman presented with hoarseness of voice, acid reflux, dysphagia and epigastric pain. Gastroscopy revealed a tumour in the lower oesophagus, which on biopsy was proved to be an invasive adenocarcinoma.

A pre-operative staging CT (computerised tomography) scan showed bilateral emphysematous changes of the lungs as well as an area of parenchymal opacity and peripheral honeycombing in the right lung apex. This was thought to represent lung scarring. All these changes were thought to represent emphysema and secondary scarring due to smoking. A PET (Positron emission tomography) scan confirmed the absence of any metastatic lesions.
The patient underwent an oesophago-gastrectomy and a small solid mass was identified within the left thoracic cavity during the surgery. This mass was situated within the pleura, between the left lung and the base of the heart. This tissue was thought to be part of a hernial sac and was excised. This lesion was not identified in the CT scan.

Three specimens were received by the histology laboratory: an oesophago-gastrectomy, two “doughnuts” and the excised “hernia sac”.

The oesophago-gastrectomy specimen was composed of a 6-cm length of oesophagus attached to part of the stomach measuring 7 cm in maximum dimension. At the gastro-oesophageal junction, 2cm from the oesophageal resection margin was a polypoid tumour measuring 3.5 cm in maximum dimension. The cut section of the tumour was solid and white in colour.

Histology revealed a moderately differentiated, invasive adenocarcinoma with large amounts of extracellular mucin. One out the four retrieved lymph nodes showed metastatic adenocarcinoma. The resection margins and the “doughnuts” were free of tumour.

The specimen which was labelled as hernia sac was an irregular portion of tissue measuring 6.0 x 4.5 x 2.0 cm. The cut section was firm with multiple small cysts ranging in size from 0.2 to 0.5cm. Microscopy revealed thick walled blood vessels and fibrotic and inflamed alveolar tissue, as well as cysts of varying sizes lined by ciliated columnar respiratory type epithelium some of which had small cartilaginous plates (Figure 1). The appearances were those of a bronchopulmonary sequestration. However, also noted within this lesion were small foci of an adenocarcinoma (figures 2 and 3). This tumour had an immunoprofile similar to the adenocarcinoma in the oesophagus. Both tumours showed strong positivity for cytokeratin 7 (CK7) and weak positivity for cytokeratin 20 (CK 20). The tumour cells were negative for thyroid transcription factor (TTF1). The histological appearance and immunoprofile confirmed that this was a metastatic adenocarcinoma arising from the oesophagus. TTF1 is a marker for thyroid and lung tissue. This stain was positive in the bronchopulmonary sequestration cells as they are of pulmonary origin, but was negative in the oesophageal adenocarcinoma cells. This contrast is highlighted in figure 3.

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**Figure 1:** Broncho-pulmonary sequestration. H&E 4x. This picture shows the cartilage and the epithelial lining of the sequestration. This is from an area not involved by the metastatic tumour.
Figure 2: Bronchopulmonary sequestration (BPS) (down) and metastatic adenocarcinoma (up) H&E stain 40x.

Figure 3: Metastatic adenocarcinoma metastasising into the bronchopulmonary sequestration. Metastatic adenocarcinoma is negative with TTF1 (up) and the bronchial epithelium (down) is positive with TTF1 which is seen as a brown colour. TTF1 immunostaining, 40x.
Discussion

Bronchopulmonary sequestrations form 0.15-6.4% of all congenital pulmonary malformations. They are usually divided into two types; intralobar or extralobar sequestration. Intralobar type is commoner than extralobar type. In Wei et al series of 2625 bronchopulmonary sequestration cases, around 84% were intralobar and 16% extralobar (3) with a ratio of around 5:1. Meanwhile, AndradeI et al quoted a ratio of 3:1 between intralobar and extralobar sequestrations in their review article (2). Bronchopulmonary foregut malformation is also considered a type of pulmonary sequestration. This is a rare anomaly in which an abnormal lung tissue is connected to the gastrointestinal tract (2).

The aetiology of pulmonary sequestration is not fully understood. There seems to be a consensus that the extralobar form is congenital, but it is still debated whether the intralobar is congenital or acquired (2). One theory suggests that the initial events leading to the formation of intralobar sequestration start with bronchial obstruction, that leads to pneumonia, local pleurisy, and parasitization of pulmonary ligament arteries (4). Review of the literature shows that the presence of neoplastic lesions in sequestration is rare. PubMed only shows nine reported cases since 1979. Two of these were squamous cell carcinomas (5, 6), two were adenocarcinomas (7, 8), three were carcinoid tumours (9, 10, 11), the other two were a blastoma (12) and a lymphoepithelioma (13). Six of the tumours arose in intralobar sequestration, two in extralobar and one in a communicating bronchopulmonary- foregut malformation. Six tumours were in men and three in women. The age range was 31-70.

Six of these tumours were located in the left lobe, five of which were in the lower segment. Two tumours were located in the right lower segment (One adenocarcinoma and one squamous cell carcinoma). The ninth case is the one originating in a communicating bronchopulmonary-foregut anomaly. The table below summarizes the main features of the previously reported cases. Case 10 in the table is the case reported in this article.

Cases of malignant tumours associated with bronchopulmonary sequestrations:

<table>
<thead>
<tr>
<th>Tumour type</th>
<th>Site</th>
<th>BPS type</th>
<th>age and sex</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 Adenocarcinoma</td>
<td>Right lower</td>
<td>EL</td>
<td>70 M</td>
</tr>
<tr>
<td>2 Adenocarcinoma</td>
<td>left lower</td>
<td>IL</td>
<td>67 M</td>
</tr>
<tr>
<td>3 Squamous cell</td>
<td>BP-FOREGUT</td>
<td></td>
<td>43 M</td>
</tr>
<tr>
<td>4 Squamous cell</td>
<td>Right lower</td>
<td>IL</td>
<td>69 M</td>
</tr>
<tr>
<td>5 Carcinoid</td>
<td>left lower</td>
<td>IL</td>
<td>41 M</td>
</tr>
<tr>
<td>6 Carcinoid</td>
<td>left lower</td>
<td>IL</td>
<td>38 F</td>
</tr>
<tr>
<td>7 Carcinoid</td>
<td>left lower</td>
<td>IL</td>
<td>45 M</td>
</tr>
<tr>
<td>8 Blastoma</td>
<td>Left upper</td>
<td>EL</td>
<td>45 F</td>
</tr>
<tr>
<td>9 Lymphoepithelioma</td>
<td>left lower</td>
<td>IL</td>
<td>31 F</td>
</tr>
<tr>
<td><strong>10 Metastatic adeno</strong></td>
<td><strong>left lower</strong></td>
<td><strong>IL</strong></td>
<td><strong>50 F</strong></td>
</tr>
</tbody>
</table>

Key: EL=extralobar, IL=intralobar, M= male, F= female, BP-foregut= communicating bronchopulmonary foregut malformation.
All the previously reported cases described primary neoplasms originating in a bronchopulmonary sequestration. This reported case is different in that it describes a metastatic tumour to a sequestration. To my knowledge this is the first documented case of a metastatic adenocarcinoma presenting within a pulmonary sequestration.

**ACKNOWLEDGMENTS**
I like to thank Mr M. Yousef, the surgeon who performed the operation and gave permission for his case to be reported. Thanks also to Dr Preethi Joseph, the histopathology registrar who performed the cut up of the case and participated in the histological diagnosis.

**References**


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