Well differentiated fetal adenocarcinoma of the lung in a 29 year old woman

K M Sheehan, J Curran, E W Kay, P Broe, A Grace

This report describes a case of well differentiated fetal adenocarcinoma of the lung in a 29 year old female smoker. The histological pattern and immunohistochemical profile were consistent with well differentiated fetal adenocarcinoma and the patient made an uneventful postoperative recovery with no recurrence after 18 months. This neoplasm is a rare lung tumour that is composed of glycogen rich neoplastic glands and tubules that resembles fetal lung at 10 to 15 weeks of gestation. It is important to identify this rare variant of adenocarcinoma because it is a low grade malignancy with low associated mortality.

Well differentiated fetal adenocarcinoma (WDFA) is a rare lung tumour that is composed of glycogen rich neoplastic glands and tubules that resembles fetal lung at 10 to 15 weeks of gestation.\(^1\) In contrast to the closely related biphasic pulmonary blastomas, the adjacent mesenchyme in WDFA is histologically benign. WDFA is a low grade malignancy and mortality from the tumour is only 15%, hence the importance of identifying this rare variant of adenocarcinoma.\(^2\)

CASE REPORT
A 29 year old woman presented with a three week history of left sided pleuritic chest pain, night sweats, poor appetite, and generalised lethargy. The patient had smoked 40 cigarettes a day for 15 years. A computed tomography scan showed a homogenous well defined mass in the medial left chest (fig 1). Bronchoscopy confirmed a left upper lobe anterior segment lesion at the orifice, which occluded the lumen.

A left upper lobectomy was performed. Gross pathological examination of the specimen revealed a single, white, 3 cm well defined mass. Low power microscopy demonstrated a proliferation of complex glands and tubules with scant stroma (fig 2). High power microscopy showed cuboidal and columnar cells, which were arranged in complex gland-like structures with cytoplasmic vacuoles (fig 3). In some areas,
the cells merged to form very small rosette-like glands, and focal areas of necrosis were identified. The intervening stroma was scant overall, and was composed of benign spindled myofibroblastic cells. In addition, squamous morules were seen (fig 4).

Periodic acid Schiff staining demonstrated glycogen within the cytoplasm of the cells. Mucin stains were negative. Immunohistochemistry showed immunoreactivity for the epithelial markers AE1/AE3 and CAM 5.2, carcinoembryonic antigen, and β-catenin, and focal positivity for epithelial membrane antigen. Oestrogen and progesterone receptors, neurone specific enolase, chromogranin, and synaptophysin were all negative. Desmin, α-fetoprotein, p53, and S-100 protein were also negative.

The histological pattern and immunohistochemical profile were consistent with WDFA. The patient made an uneventful postoperative recovery with no recurrence after 18 months.

DISCUSSION
The most recent classification by the World Health Organisation in 1999 has removed WDFA tumours from the pulmonary blastoma category and classified them as a variant of adenocarcinoma. Review of the literature reveals several classic features of WDFA, all of which were seen in our case. The most recent paper on WDFA describes aberrant cytoplasmic expression of β-catenin' in these tumours, which was also seen in our case.

**Take home messages**

- Well differentiated fetal adenocarcinoma of the lung is a rare tumour that is composed of glycogen rich neoplastic glands and tubules that resembles fetal lung at 10 to 15 weeks gestation
- It is important to identify this rare variant of adenocarcinoma because it is a low grade malignancy with low associated mortality

“Pulmonary blastomas show similar fetal-type malignant glands but also contain an embryonic malignant stroma”

The differential diagnosis of this tumour includes high grade adenocarcinoma of fetal lung type or clear cell adenocarcinoma with fetal lung features. The well differentiated appearance and lack of pleomorphism may suggest carcinoid yet the morules and the lack of chromogranin immunoreactivity are inconsistent. Finally, pulmonary blastomas show similar fetal-type malignant glands but also contain an embryonic malignant stroma.

References

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