Pulmonary acinic cell carcinoma (Fechner tumour) with carcinoid component. A case report

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The authors present a very rare case of primary lung acinic cell carcinoma with carcinoid component in a 53-year-old man.

Key words: rare pulmonary neoplasm, Fechner tumour, acinic cell tumour, lung carcinoid.

Among bronchial tumours connected with bronchial glands we distinguish mucoepidermoid carcinoma, adenoid cystic carcinoma, epithelial-myoepithelial carcinoma, pleomorphic adenoma and acinic cell carcinoma. The last two are extremely rare. The first case of acinic cell carcinoma was described by Fechner in the year 1972 [1]. Since then there have been only a few new cases described. Moran *et al.* [2] have published the largest group of such cases. Tumours formed from two components, acinic cells and neuroendocrine carcinoid cells, are even more rare. In the recent English literature there are two reports concerning mixed carcinoid and acinic cell tumours from Japan and Italy [3, 4]. The rarity of this kind of tumours may lead to incorrect diagnosis when pathologists first come across such cases. Recently we have had an opportunity to diagnose one tumour of this type.

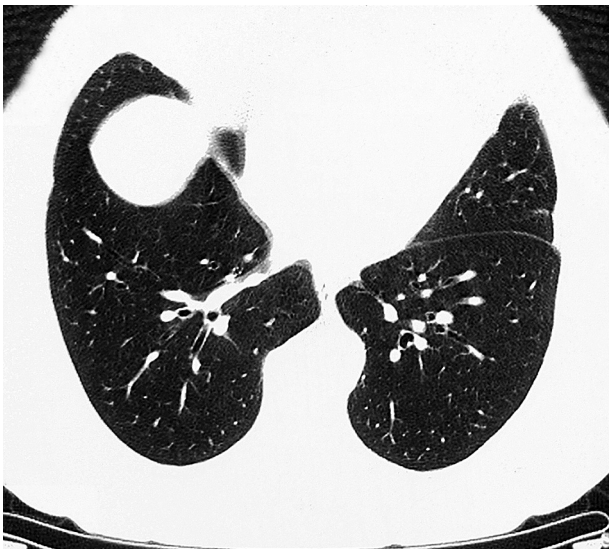

Fig. 1. CT scan shows large tumour in the right lung

Material and methods

A 63-year-old man was admitted to the hospital in January 2009 because of a tumour in his right lung in the vicinity of the mediastinum visible on chest X-ray (Fig. 1). There were no other signs of the disease. During surgery the tumour was removed together with part of the upper and middle right lobes. Intraoperative pathological study suggested a neoplastic tumour – probably liposarcoma. The tumour was 52 × 23 × 18 mm in diameter. Material was fixed in formalin and embedded in paraffin blocks No. 190019. Frozen material was used for Sudan III staining. Slides and paraffin blocks were sent for consultation to the Chair of Pathomorphology, Jagiellonian University Medical College. The slides from paraffin blocks were used for HE staining, PAS with/without previous diastase digestion, Gomori silver staining as well as immunohistochemical staining against: cytokeratin AE1/AE3, cytokeratin 5/6, chromogranin A, synaptophysin, S-100, EMA, α1-antichymotrypsin, smooth muscle actin, CD10, Ki67, CD68, CD99, HMB45, lysozyme, calretinin, and NSE. Additionally, formalin-fixed, paraffin embedded tissue slides

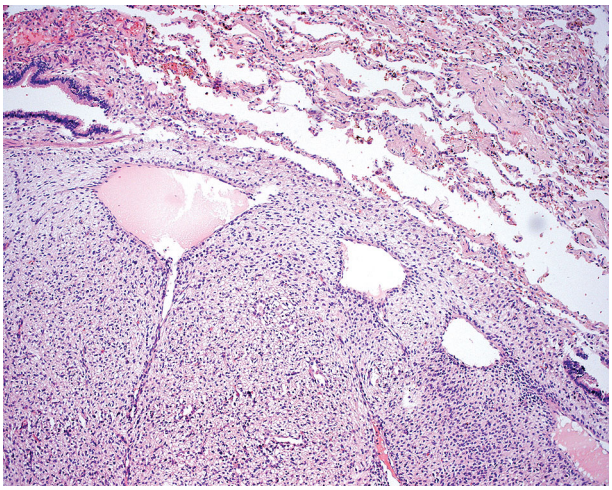

Fig. 2. Border of the lung tumour. Tumour cells have clear cytoplasm and centrally located nuclei

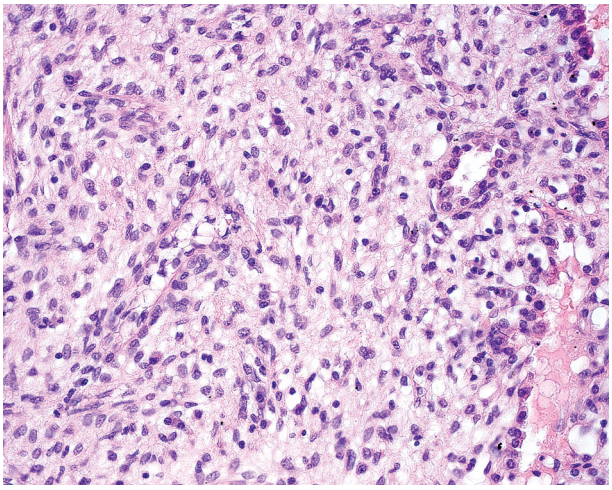

Fig. 3. In some parts of the tumour there are clefts and glands lined by epithelial cells

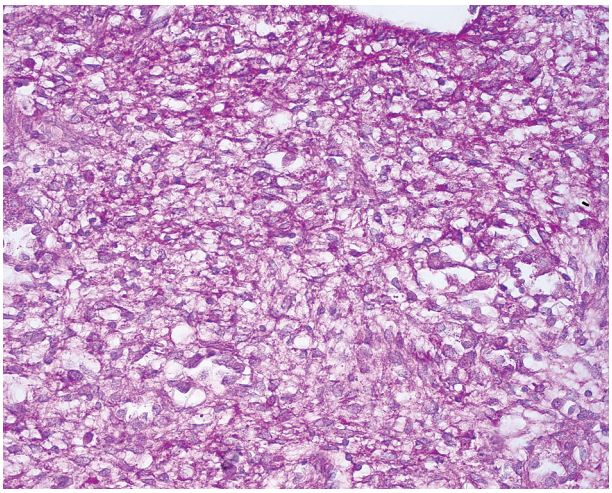

Fig. 4. PAS staining strongly positive in cytoplasm of the tumour cells

were used for FISH analysis with commercially available *SYT* Dual Color Break Apart probes (Vysis LSI SS18 LOT 420 582).

Results

Histological analysis revealed a neoplastic tumour with solid growth pattern, composed predominantly of a sheet of cells with clear, empty cytoplasm and distinct borders (Fig. 2 and 3). Both PAS and PAS treated with diastase staining were distinctly positive (Fig. 4) but Sudan III was negative in their cytoplasm. Nuclei of the cells were eccentrically located, round or oval, and only a few cells presented visible nucleoli. Some cells looked like signet ring cells. No mitotic figures were seen, but 20 to 30% of nuclei were positively stained with Ki67 antibody. At the border, the tumour infiltrated the walls of smaller bronchi. In the central part of the tumour, there were many thin-walled, mostly capillary blood vessels. They were focally surrounded by lymphocytic infiltration. Clear cells were stained negatively for EMA and cytokeratins; a few of them were dyed immunohistochemically for lysozyme.

On the border of the tumour and less commonly in central parts there were dispersed tubular and slit-like structures lined by cuboidal cells with a small amount of cytoplasm; all these cells were stained with antibodies against pancytokeratin (Fig. 5), EMA as well as for chromogranin (Fig. 6) and synaptophysin. Antigens HMB45, CD68, CD10, S-100, cytokeratin 6/8 and calretinin were negative, smooth muscle actin positive only in the wall of small blood vessels. CD99 was weakly positive in parts epithelioid cells lined tubular structures.

Since the surgery in January 2009 the patient has remained alive with no signs of the disease.

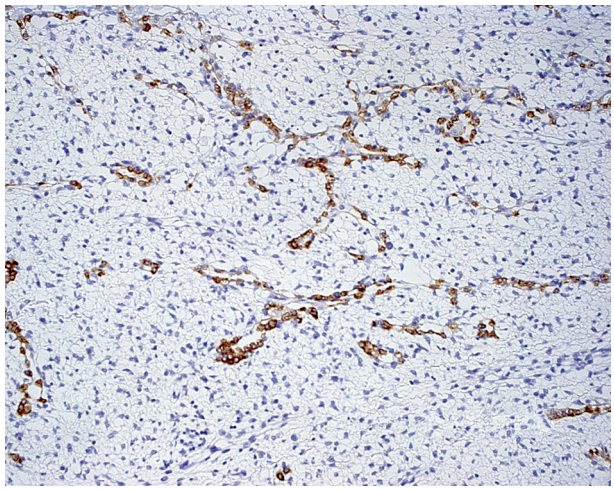

Fig. 5. Immunohistochemical staining for pan-cytokeratin positive in epithelial cells lining glands and clefts

Discussion

The first step in differential diagnosis of this tumour is clinical investigation and exclusion of metastases of clear cell carcinoma from the kidney and adenocarcinoma from salivary gland.

Primary bronchial squamous cell carcinoma and adenocarcinoma with clear cells have many typical features which enable the differentiation.

Other possible diagnoses are sugar tumour – clear cell tumour (pecoma) and clear cell carcinoid. Sugar tumour, also known as clear cell tumour, is characterized by a positive reaction with HMB45 and S-100. These antigens were negative in the presented tumour. Clear cell carcinoid is a very rare type of carcinoid tumours and description of the first case in lung was published in 1998 by Gaffey *et al.* [5]. All cells in clear cell carcinoid are stained positively with antibodies against chromogranin, synaptophysin and neuron-specific enolase. These cells are negative with fat staining and PAS staining. Also electron microscopy investigation shows characteristic neurosecretory-type granules in the cytoplasm. Our tumour presents partially neuroendocrine differentiation but clear cells do not look like typical carcinoid cells because of negativity for chromogranin and synaptophysin and the positive PAS staining. Rodriguez *et al.* [4] reinforced the hypothesis that both neuroendocrine and acinic cells derive from a common stem cell. It is possible that some carcinoids and acinic cell carcinomas located in lungs are mixed tumours but were not examined as such.

Another problem is differentiation of this tumour with very rare biphasic synovial sarcoma with myxoid stroma [6]. Negative investigation both for specific breaks of chromosome 18q11 and calretinin expression makes it possible to reject the diagnosis of synovial sarcoma.

Lack of cytokeratin and EMA staining in clear cells is observed in many cases of clear cell carcinoma of salivary glands [7].

Prognosis for a patient with acinic cell carcinoma after surgery is rather good. A few cases with local recurrences and metastases to the lymph nodes have been described so far [8, 9].

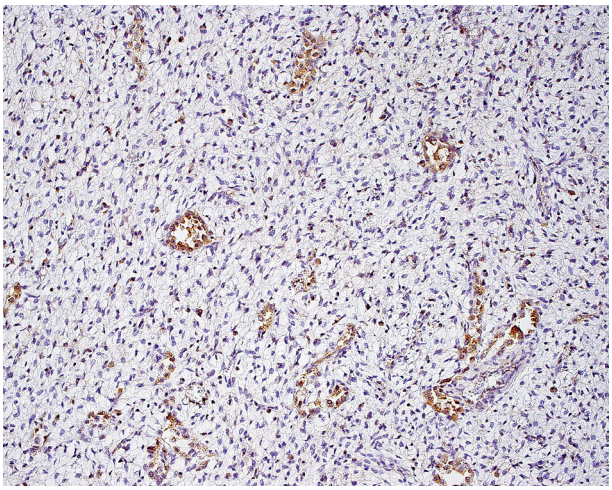

Fig. 6. These epithelial cells are also positive for chromogranin

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