KARACİĞER METASTAZI İLE SEYREDEN PRİMER AKCİĞER ADENOİD KİSTİK KARSİNOMU; ASTIM İLE İZLENEN OLGU

PRIMARY PULMONARY ADENOID CYSTIC CARCINOMA WITH LIVER METASTASIS: FOLLOWED BY DIAGNOSIS OF ASTHMA

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ÖZ

Adenoid kistik karsinom (AKK) adenokanserlerin bir varyantı olarak kabul edilir ve farklı histopatolojik ve klinik özelliklere sahiptir. AKK çoğunlukla tükrük bezlerinden kaynaklanır. Daha nadir olarak da meme, deri, serviks, üst sindirim sistemi ve akciğerden de köken aldığı görülmüştür. Akciğerin primer AKK'sı nadir görülür ve tüm akciğer kanserlerinin %0.04-0.2'sidir. Klinikte pulmoner AKK'nın nadir bildirilmesinin yanında uzak metastazı olanlar daha da nadir olarak bildirilmiştir. 63 yaşında bayan hasta 3 yıldır astım tanısı olup son bir aydır nefes darlığı, öksürük, balgam yakınmalarında artış olmuş. Çekilen toraks bilgisayarlı tomografisinde (BT) sağ ana bronşu dıştan bası ile daraltan yumuşak doku dansitesi, sağ alt lobda konsolidasyon-obstrüktif pnömoni ve sağ plevral sıvı bulguları izlenmesi üzerine ileri tetkik ve tedavi için hastanemize yönlendirilmiş. Bronkoskopi yapılan olgunun transbronşiyal ince iğne aspirasyon biyopsi (TBİİAB) sitolojisi malign olup endobronşiyal biyopsi AKK olarak geldi. Karaciğer parankiminde en büyüğü yaklaşık 7cm çaplı birbiri ile birleşme eğilimindeki hipodens lezyonlarda artmış florodeoksi glukoz (FDG) tutulumu (SUVmax: 10.3) mevcuttu. Evre 4 olması nedeniyle hastaya cerrahi düşünülemedi, kemoterapi başlandı. AKK genellikle yavaş büyüyen bir tümör olarak bilinmekle birlikte hızla yayılabilir. Olguyu astım tanısı koymadan önce detaylı incelemenin yapılması gerekliliğine ve AKK tanısındaki gecikmenin hastalığın ilerlemesinde ve ileri evrelere geçmesine neden olabileceğine dikkat çekmek için sunmaya uygun bulduk.

ABSTRACT

Adenoid cystic carcinoma (ACC) is considered as a variant of adenocarcinomas and has different histopathological and clinical features. ACC is mostly originated from salivary glands. It can be originated from rarely, breast, skin, cervix, upper gastrointestinal system and lung. Primary ACC of the lung is rare and 0.04-0.2% of all lung cancers. In addition to the rare presentation of pulmonary ACC, patients w ith distant metastases have been reported rarely. A 63-year-old female patient has been followed with the diagnosis of asthma for 3 years. She had dyspnea, cough, and increased sputum complaints for the last one month. On the chest computed tomography (CT), soft tissue density narrowing the right main bronchus, consolidation –obstructive pneumonia on the right low er lobe and right pleural fluid findings w ere monitored and the patient was referred to our hospital for further examination and treatment. Fiberoptic bronchoscopy (FOB) was performed and transbronchial needle aspiration biopsy (TBNAB) was malign and endobronchial biopsy revealed ACC. Increased Fluoro Deoxy Glucose (FDG) uptake (SUVmax: 10.3) was present in hypodense lesion that tended to merge with 7 cm diameter in the liver on PET-CT. Patient staged as T3N2M1- Stage 4. Chemotherapy was initiated, surgery could not be considered. ACC is generally known as a slow-growing tumor, but it may spread rapidly. We presented the case to draw attention to the necessity of adequate examination of the patient before the diagnosis of asthma and delay in the diagnosis of ACC cause the disease to progress to advanced stages.

INTRODUCTION

Adenoid cystic carcinoma (ACC) is considered as a variant of adenocarcinomas and has different histopathological and clinical features. It is thought to be related to proliferation of myoepithelial and secretory epithelial cells. But its occurence is still not fully understood (1). ACC is mostly originated from salivary glands. It can be originated from rarely, breast, skin, cervix, upper gastrointestinal system and lung. All of these ACCs exhibit similar histologic features regardless of where their primary site (2).

Primary ACC of the lung is rare and 0.04-0.2% of all lung cancers. In addition to the rare presentation of pulmonary ACC, patients with distant metastases have been reported rarely. Slow growth, long life expectancy, asthma or recurrent pneumonia are the hallmarks of the clinic (3). The case of ACC with distant metastasis was presented with the literature.

CASE

A 63-year-old female patient have the diagnosis of asthma for 3 years. She had irregular asthma treatment. When the patient history of asthma deepened it was tought that there was no asthma. Her dsypnea was increased, cough and sputum were added for the last month. On the chest x-ray (Figure 1) she had hilar enlargement and consolidation and atelectasis findings on the right lung. On the chest computed tomography (Figure 2a, 2b), soft tissue density narrowing the right main bronchus, consolidation-obstructive pneumonia on the right lower lobe and right pleural fluid findings were monitored and the patient was referred to our hospital for further examination and treatment.

On bronchoscopy, right main bronchus was compressed from the outside and obstructed with polypoid tissue. Transbronchial needle aspiration biopsi (TBNA) and forceps biopsies (FB) were performed. The cytology was malignant and endobronchial biopsy revealed ACC (Figure 3a). Immunohistochemical SMA (+), S-100 (+), p63 (+), CK7 (+), TTF-1 (-) (Figure 3b). The cranial MR scan was normal. There was a 3.2 cm soft tissue lesion extending to the subcarinal area accompanied by atelectatic, consolidative changes in the periphery surrounding the right main bronchus and increased FDG in the subcarinal lymph node (SUVmax: 5.5), increased FDG uptake in the consolidation area (SUVmax: 6.7) involving the left lower lobe showed FDG uptake in the same level as surrounding tissue, increased FDG uptake (SUVmax: 10.3) was present in hypodense lesion that tended to merge with 7 cm diameter in the liver on PET-CT (Figure 4a, 4b). Patient staged as T3N2M1- Stage 4. Cisplatin-gemcitabine chemotherapy was initiated due to ACC. After 4 cycles of cisplatin-gemcitabine, progression was observed. Paclitaxel was initiated as the second line chemotherapy. The patient is stable and is stil under treatment.

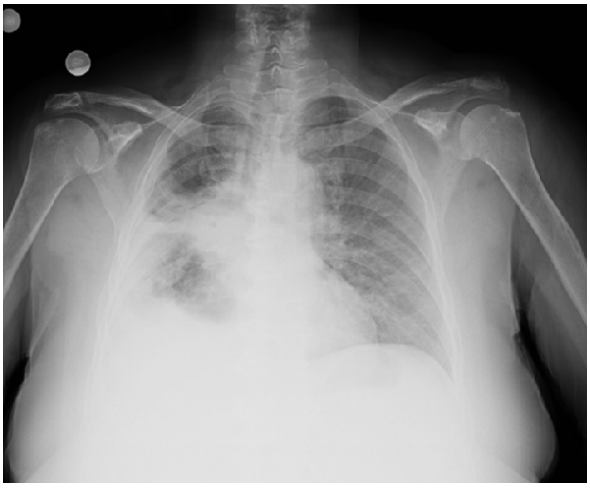

Figure 1. Hilar enlargement, consolidation and atelectasis findings at the right lung on chest X-ray

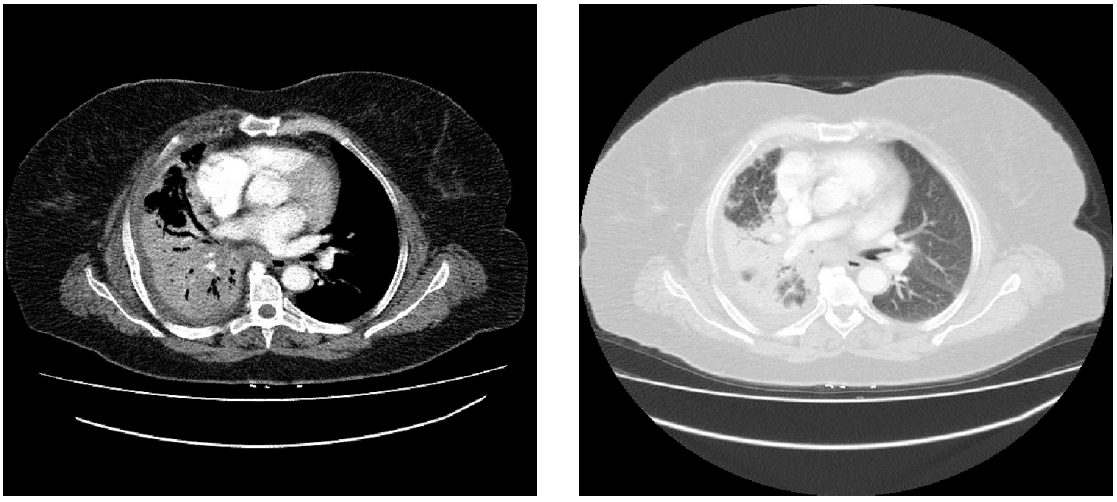

Figure 2. Soft tissue narrowing the right main bronchus, consolidation-obstructive pneumonia, pleural fluid at the right lung on CT

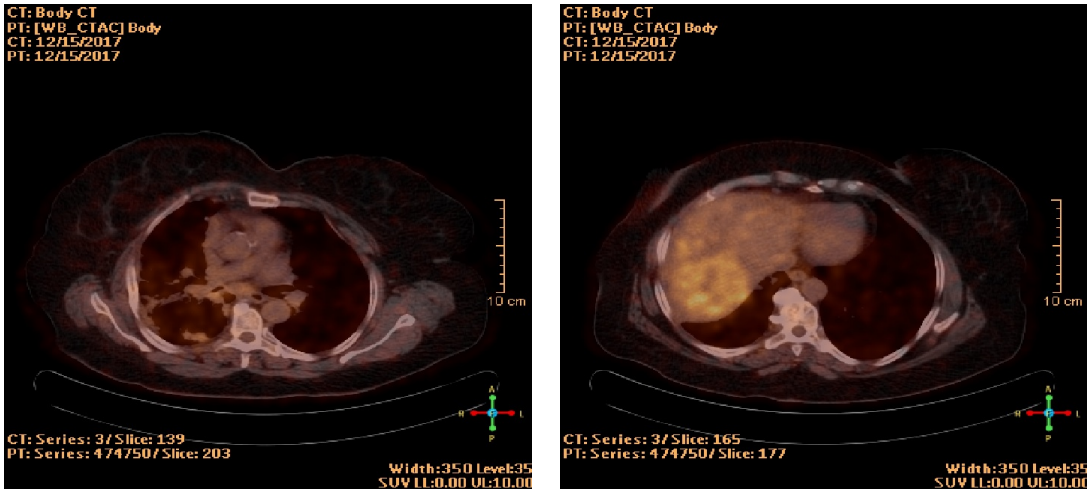

Figure 3. PET- CT scan showing increased FDG uptake in hypodense lesions

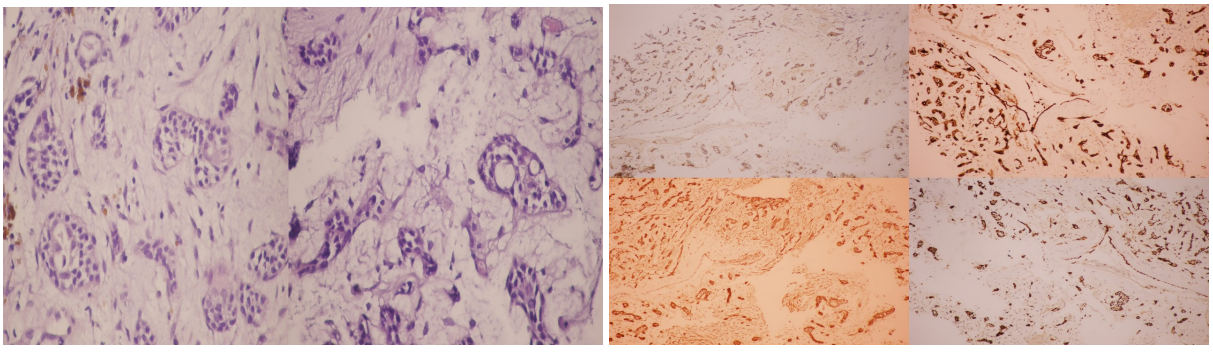

Figure 4a. Endobronchial biopsy revealed ACC with hematoxsylene-eosin staining **b.** Immunohistochemical SMA (+), S-100 (+), p63 (+), CK7 (+)

DISCUSSION

It was first described in 1859 by the name of Billroth silindroma, but nowadays the term, adenoid cystic carcinoma is used (4). It is mostly seen in both sexes around fifty years. ACC is most commonly seen in the major and minor salivary glands and seen in the head and neck region, but rarely in the lung, breast, cervix, upper gastrointestinal tract and skin (5). ACCs of organs other than salivary glands are rare tumors and have histological features similar to ACCs of salivary glands. Therefore, the clinicopathological features are thought to be similar to those of salivary gland origin. However, breast ACCs have a better prognosis than salivary gland ACC (6). The prognosis of esophageal ACC is worse (7). ACC's behavior is not fully understood. It is originated from the submucosal bronchial secretory glands in the lung and is most commonly seen in the trachea and main bronchi. As the number of secretory bronchial glands decreases as going to the distal bronchi, the incidence decreases. It occurs in approximately 0.04-0.2% of all primary lung cancers (8). It is a slow growth tendency with rare occurrence, and life expectancy is long.

The patient had been suffering from asthma for 3 years because of shortness of breath, cough and wheezing. After the lesion was visible on the chest radiography, it could be diagnosed and diagnosed as ACC. Therefore, in patients with such complaints, chest X-rays should be evaluated well before the diagnosis of asthma, and CT should be performed in the required cases.

Optimal treatment for this disease is surgical resection, if possible. In the past, the prognosis was quite bad, and in 5 years life expectancy increased to 60-100% in cases operated with advances in surgical techniques. However, many patients are not candidates for surgery and although the tumor is resected, the risk of local recurrence is high due to incomplete resection. Cases reported in which postoperative radiotherapy provide the control of residual tumor and provide contribution to survival (9). Gedik et al. reported that they applied 64 gray curative radiotherapy to a patient who could not be operated due to the capacity of lung and achieved a decrease in the size of the mass and obstruction. (10). Because our case was stage 4, radical RT could not be considered, and palliative RT could be tried if there was a need for palliation.

Unlike other lung cancers, the residual tumor tends to grow slowly and life expectancy is long. 2/3 of tracheabronchial adenoid cystic carcinoma is seen in the trachea and the remaining in the main bronchus and distal.

AKK is a malignant tumor and can metastasize. Metastasis can be found in regional lymph nodes in 10-20% of newly diagnosed ACC (11). Distant metastasis can be seen in the late stages of the disease and usually lung, bone, liver and brain metastases are observed (12). Our patient had liver metastasis at the time of diagnosis. Therefore, surgery could not be considered. Although it is not clear whether chemotherapy will alter the natural course of metastatic ACC, chemotherapy was initiated as stage 4 disease in our case. In the first line, progression was monitored and the second line was started. Studies on the use of targeted therapies in ACCs are ongoing. In the literature, a case of lung ACC with a positive EGFR mutation and a successful use of tyrosine kinase inhibitors has been reported (13). Although carcinomas other than lungs have not been reported to be EGFR positive, there are several studies investigating EGFR mutations in ACC. In our case, we could not look for EGFR mutation.

Pulmonary ACC may be more aggressive than those of salivary gland origin. Therefore, it may be necessary to start more aggressive treatments such as radiotherapy and chemotherapy early. Because of this difference in prognosis, important prognostic factors should be determined in pulmonary ACCs.

The prognostic value of the histological stage of the tumor is contradictory between salivary glands and other ACCs. The growth pattern of the histological universe and its association with prognosis have been reported (1). The possible relationship was also shown between histological stage and treatment results (14). Moran et al. reported that the histological features at the time of diagnosis, but not the histological stage, would determine the clinical outcome. Life-time relationship with lymph node involvement is not reported respectively (11,12).

There is no study showing which organ is involved in distant metastasis and its relation to prognosis. The presence of bone metastases in head and neck ACCs have been associated with a potentially poor prognosis, with a median survival of 21 months after bone metastasis and 54 months after lung metastasis (15). When the prognosis is compared to the metastatic organs, bone metastasis is more resistant to chemotherapy and fast progressive. It is also associated with poor prognosis for lungs. Liver metastasis was present in our case. There was no information about the relationship between liver metastasis and prognosis in the literature.

CONCLUSION

ACC is generally known as a slow-growing tumor, but it may spread rapidly. Therefore, factors that determine the aggressive behavior of the tumor should be determined and individual and organ specific treatments should be planned. We presented the case to draw attention to the necessity of adequate examination of the patient before the diagnosis of asthma and delay in the diagnosis of ACC cause the disease to progress to advanced stages.

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