A rare association; pulmonary blastoma developing on extrapulmonary sequestration basis

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Abstract

Pulmonary blastoma (PB) is a thoracopulmonary mesenchymal disembyogenic neoplasm which is rarely seen and generally in childhood. Pulmonary sequestration is one of the less observed congenital malformations. A 45-year-old female patient who was diagnosed with PB, histopathologically developed on atypically placed extrapulmonary sequestration in the left upper zone is presented in this study. Pulmonary sequestration is one of the less observed congenital malformations. It includes 0.15–6.4% of all congenital lesions [3, 4]. It is mostly (90%) observed on the left lung near to the basal area. Its arterial supply takes root mostly from the thoracic aorta whereas the others are from abdominal aorta.

In this paper, we present a female patient who was histopathologically proven to have PB, which had developed on a rarely placed ES basis. It was discussed in accompany with literature due to its rare presentation and poor prognosis.

2. Case presentation

A 45-year-old female patient presented to our clinic with chest and back pain, asthenia, cough, dyspnea and headache commencing in the last two months. BP: 110/70 mmHg, RR: 19/min, fever: 36.7 °C were found. In physical examination, there was no pathological finding except for a decrease in respiratory sounds in the left lung left zone. ECG was in normal sinus rhythm, blood biochemistry and complete blood count (CBC) examinations were normal. In PA lung graphy, opacity and sinus blunting were detected in the left lung upper zone. In thorax computerized tomography (CT), a 4×4 cm sized mass in the para mediastinal area adjacent to the upper lobe apical segment of the left lung and pleural fluid in left median and lower zone were monitored (Fig. 1a). Pleural fluid examination: it was determined as exudate, no malignancy finding was observed in histopathological examination. PET-CT was requested with a suspicion of a possible malignancy. In PET-CT-scan: a mass that in the middle is hypometabolic and sides are hypermetabolic, and to be considered as visseral pleura invasion in the left lung upper lobe apical segment [standard uptake value (SUV): 9,1]. Pleural effusion which presents hypermetabolic involvement in apicoposterior segment (SUV: 3,3) was monitored (Fig. 1b).

No pathological finding was detected except for external compression in the left upper lobe bronchus in the bronchoscopy performed. In bronchoalveolar lavage fluid, no malignancy was detected in histopathological examination and no ARB was observed in microbiological examination.

Benign cytology was reported in pleura biopsy performed. The patient was operated at first in elective conditions for mediastinal malignancy. We performed a left posterolateral thoracotomy and observed an extrapulmonary mass of 8×12 cm diameter. Intraoperatively we saw a non-functioning and calcified bronchus of the mass opening into trachea and its vein draining into main pulmonary vein (Fig. 1c). We could not find an arterial structure except a small diameter artery in association with subclavian artery in the apikal part of the mass. Artery and vein were tied and cut. Bronchus was cut and closed; the mass was excised (Fig. 1d). No problem arose after the mass excision and the patient was discharged on postoperative day 7. Histopathological examination of the material was reported complying with PB (Fig. 2a,b).
In 60% of the cases in the first decade of life this could be due to infection and trauma. While the diagnosis of ES can be done preoperatively, it occurred as the result of the operation. Nevertheless even in the literature the possibility of a preoperative diagnosis of an ES case is reported to be only 6% [6].

PB is a rare malignancy with poor prognosis and it is frequently observed in cystic lung pathologies [7]. Even though the PB etiology is unknown, some authors propose that there is a relationship between smoking and blastoma [8]. PB may be seen with congenital diseases and could possibly be hereditary. PB association with congenital lobar emphysema and neurofibromatosis was reported [9]. Our case is a middle-aged female patient with no other disease. No smoking and congenital disease were present.

There are no cases in literature for this association. Probably malignity due to ES is a possible occasion. PB developing on an ES basis is very rare and probably a coincidental association. We suggest that malignancies should be searched unless they are rare besides degenerations in ES.

Although chemotherapy and radiotherapy following radical surgical resection are the treatment options, the prognosis is very poor. It was reported that nodal involvement is the most important indicator in PB [10]. No intraoperative lymph node was detected in the patient and chemotherapy application continued after the surgical resection.

As there was no lymph node involvement on the preoperative thorax CT scan, we did not observe any nodes during the operation. After ten months of follow-up we detected no symptom or sign of locoregional relapse and she is disease free and healthy under regular control.

We did not detect ES and PB association in the literature. We wanted to share this rare case. We consider that malignancies should be searched unless they are rare besides degenerations in ES.

3. Conclusion

Although sequestrations are asymptomatic, they may become symptomatic by events such as infection, degeneration and trauma. While the diagnosis of ES can be done in 60% of the cases in the first decade of life this could be achieved for only half of the patients with IS cases [3]. Several ES cases existing in the right and left hemithorax upper zones are cited in the literature [5]. Initially a diagnosis was done in our patient as a mediastinal malignancy but we never thought about ES at the same time but it occurred as the result of the operation. Nevertheless even in the literature the possibility of a preoperative diagnosis of an ES case is reported to be only 6% [6].

References