A case report of Pulmonary blastoma

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Abstract:
Pulmonary blastoma is a rare tumor of the lung. It is a mixed tumor containing malignant epithelial and connective tissue elements. It usually presents late as a large tumor in the periphery of the lung. The tumor has a poor prognosis. The treatment of choice is surgical excision. Adjuvant chemotherapy and radiotherapy is given for metastatic disease. We present a case of pulmonary blastoma and the literature is reviewed.

Keyword: Pulmonary blastoma, lung malignancy

A 42 year old lady of south indian origin was refered by the pulmonologist to the cardiothoracic surgery out patient department for management of a mass which was located in the upper lobe of the right lung noted in the computerised tomography (CT) thorax. She complained of dry cough for 6 months. Mantoux test was negative. Sputum for AFB was negative. No radiological change was noted after a course of antibiotics. Chest X ray showed a round opacity in the right upper zone.

Figure 1 Chest X ray showing a round opacity in the right upper zone

(Figure 1). CT chest showed a round mass located in the left upper lobe of size 10x6 cms(Figure 2).

Figure 2. CT chest showing a right upper lobe mass
CT abdomen was normal. Bronchoscopy was normal. All her blood investigations were within normal limits.

Patient underwent right posterolateral thoracotomy under general anesthesia. On examination of the right upper lobe, a tumor of size 10x7 cms which was firm in consistency and well defined was noted(Figure 3).

Figure 3. Intraoperative picture of the tumor in thright upper lobe

There was no lymphnode enlargement. The rest of the right lung appeared normal. Right upper lobectomy was done. Her postoperative course and convalescence were normal.

Surgical specimen showed a tumor which was firm in consistency (Figure 4)

Figure 4. Gross specimen showing the upper lobe of right lung and the tumor. Cut section shows areas of hemorrhage and necrosis

The tumor was 10x7 cms in size. The tumor was located subpleuraly and the surrounding lung tissue was compressed by the tumor. Microscopically the tumor consisted of numerous branching ducts and clefts lined by columnar and cuboidal epithelial cells (Figure 5)

Figure 5. Histology showing branching ducts and clefts lined by columnar and cuboidal epithelial cells

The cells were well differentiated. These epithelial cells were embedded in cellular stroma containing spindle cells and myxomatous tissues containing few smooth
muscle fibers. The cells were regular and showed few mitotic figures. The features were suggestive of pulmonary blastoma. Pulmonary blastoma is an interesting tumor due to its controversial histological characteristics. Originally it was called an embryoma by Barnard. It was renamed a blastoma by Spencer based on the mesodermal origin of both epithelial and stromal components of the lung. Pulmonary blastoma can be benign or malignant. The macroscopic appearance of the tumor is that of a well defined, well demarcated tumor located subpleural in the periphery of the lung. It is a solid tumor with no bronchial connections. Malignant tumors are also well demarcated by surrounding fibrous lung tissue. The cut surface of the tumor will show a white or grey tumor with areas of granular, friable, hemorrhagic and necrotic areas. The epithelium lined tubes or tubules are similar to that seen in fetal lung tissue. These tubes are surrounded by primitive looking mesenchymal stroma which also looks neoplastic. They differentiate into fibrocytic tissue, muscles and cartilage. The epithelial lined cleft end in solid nests of polygonal cells. Pulmonary blastoma presents within the age group of 2 months to 70 years. Incidence is more common in the age group of 40 and 60 years. Males are affected nearly 3 times as frequent as females. The most common presenting symptoms are cough and hemoptysis. The primary treatment of choice for pulmonary blastoma is surgery. Adjuvant chemotherapy and radiotherapy is the mode of treatment in patients with metastatic disease. Surgical treatment for pulmonary blastoma include pneumonectomy, lobectomy and segmental resections. Chemotherapy is given with combination of drugs like vincristine, cyclophosphamide, dactinomycin or doxorubicin. Pulmonary blastoma is recognised as a distinctive tumor. These tumors are more similar to carcino sarcoma than to true blastoma. These tumors have unique pathological, clinical and prognostic features, hence they are recognized as distinct neoplasm.