



Pulmonary Blastoma with Germ Cell Differentiation with Pancytopenia- A Rare Case with Even Rarer Presentation

Authors

Dr Milap Shah, Dr Priyanka Telkar, Dr Sachin Johari, Dr Ravindra Vottery

Dr Harikishan

Yashoda Hospital

Abstract

Pulmonary blastoma is a rare lung neoplasm of disputed histogenesis and variable biologic behaviour. A 31 yr old male presented to the hospital with complaints of breathlessness. Imaging studies revealed a mediastinal mass measuring 11 x 9 x 7cm. Subsequent histopathology on core biopsy revealed a biphasic tumor showing mesenchymal and epithelial components. It is a rare tumor with aggressive behaviour. The case is being presented to make the general histopathologists aware of this rare entity.

Introduction

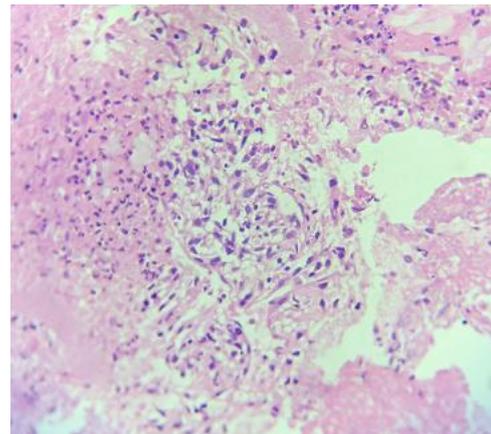
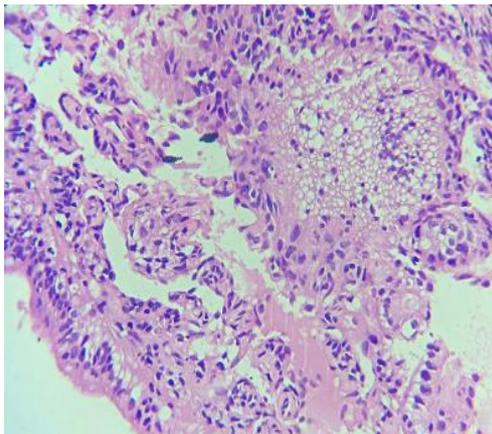
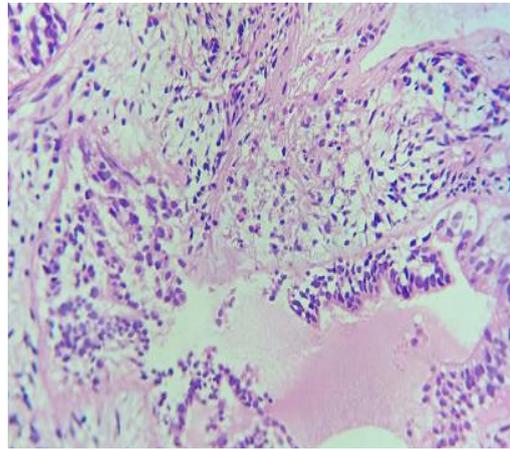
Pulmonary blastoma is a rare lung neoplasm comprising 0.25-0.5% of all primary lung tumors and portends a poor prognosis. Typical cases contain both epithelial and mesenchymal tissues and a variety of patterns have been described but the expression of oncofetal antigens in these tumors has been noted rarely. NO such case has been reported so far in PubMed indexed journals with bone marrow involvement.

Case Report

A 31 year old male presented to pulmonology opd with history of breathlessness since 1 month. There were no other complaints. He had no significant past history. Routine hemogram revealed marked pancytopenia with with platelet count as low as 4000cells/dl. Chest xray showed homogenous opacification of right mid zone and lower zone, suggesting effusion with underlying

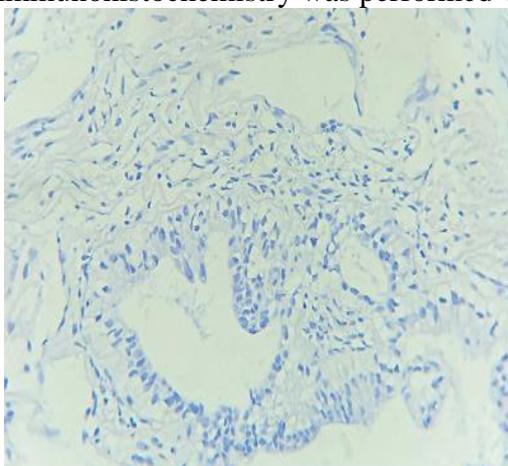
consolidation. Rest of the lung parenchyma appeared normal. Further imaging studies revealed a huge right anterior mediastinal mass measuring about 11x 9x 7 cm. Investigations showed raised Lactate dehydrogenase (LDH), Alpha-fetoprotein (AFP) and B-HCG. Based on the above findings ultrasound guided core biopsy was taken and histopathology revealed the following findings.

Biopsy cores showed a biphasic tumour showing mesenchymal and epithelial elements. Epithelial elements were in the form of glands which show nuclei pushed towards the lumen. Stroma showed spindle cells with variable edema and primitive at places. A diagnosis of primitive tumour with biphasic pattern was given. Differentials being considered were Pulmonary blastoma with Germ cell differentiation and Mixed Germ cell tumour with primitive stroma of teratoma.

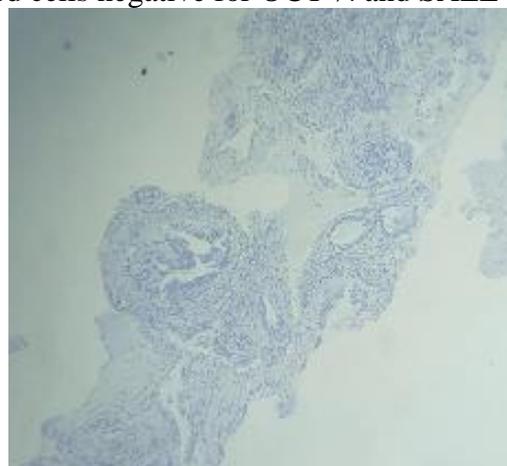


HE 40x showing epithelial component and mesenchymal component

Immunohistochemistry was performed which showed cells negative for OCT $\frac{3}{4}$ and SALL4.



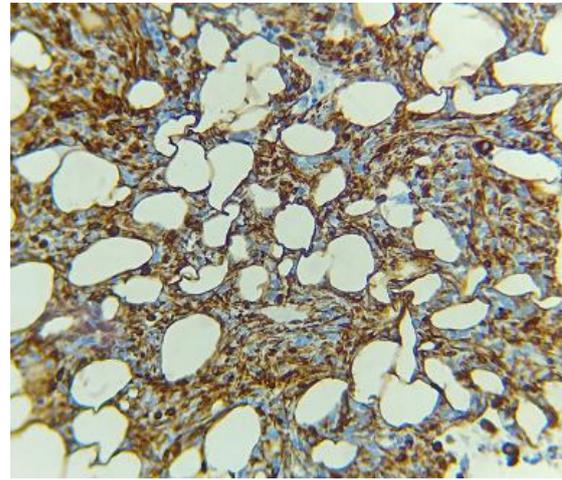
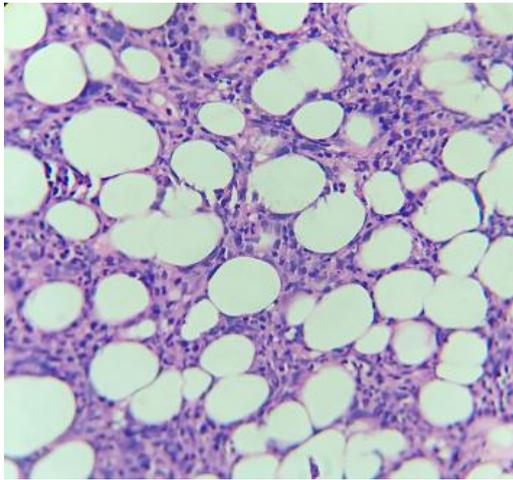
IHC: OCT $\frac{3}{4}$



IHC: SALL4

PET CT scan was performed which showed increased uptake in the bone. Bone marrow biopsy revealed increased marrow cellularity for the age and prominence of abnormal round to oval cells along with spindle cells. The cells had coarse chromatin with prominent nucleoli in few of them. Report was consistent with High grade tumour deposits. Immunohistochemistry was performed which showed abnormal neoplastic cells strongly

and diffusely positive for Vimentin. These neoplastic cells were negative for LCA,CK, CD3, CD30, TDT, CD34, SALL4, OCT $\frac{3}{4}$, Myogenin, WT1, PAX5, CD23 and MPO. Based on the above findings a diagnosis of bone marrow involved by Undifferentiated primitive spindle cell/Sarcomatous component of Pulmonary blastoma were made.



Bone marrow biopsy showing interstitial infiltration by primitive cells positive only for Vimentin

Discussion

The first case of Pulmonary blastoma was described by Barnett and Barnard 1945. Since then only about 300 cases have been reported worldwide. The cancer was named by Spencer for the similarity of its microscopy to the fetal lung at 10-16 week stage of development. Pulmonary blastoma is an uncommon lung malignancy, usually presenting itself as a large chest mass causing pain, hemoptysis, cough and dyspnea. However, it is asymptomatic in upto 40% of the cases. It is a very aggressive form of lung carcinoma that contains the presence of two main components that include fetal adenocarcinoma and primitive mesenchymal stroma. Hence it is known as a biphasic tumour. The mean age of presentation being 39-53 years, 20% occurring in patients less than 20 years. According to WHO, a definitive diagnosis of pulmonary blastoma can only be made on a surgically-biopsied tumour specimen.

Fine needle aspiration cytology of the primary diagnosis is usually not helpful in reaching a diagnosis. Image guided biopsies when planned should yield multiple needled cores keeping in view the characteristic morphological diversity of the tumour. A single needle biopsy core may sample a single tissue type with relatively bland nuclear features, hence may conceal the true nature of the disease.

Microscopically, the epithelial and mesenchymal components have a primitive “fetal-type”

appearance. The well formed tubular glands are surrounded by cellular stroma of embryonal appearance, which resembles Wilm’s tumour or fetal lung of 10-16 weeks. Glandular cells are tall, columnar, often with clear cytoplasm and subnuclear and supranuclear cytoplasmic vacuoles. Stroma may differentiate towards striated muscle, smooth muscle or cartilage.

Conclusion

Pulmonary blastoma is a rare and malignant neoplasm with poor prognosis. It grows rapidly, and diagnosis is problematic due to its two-phase construction and localization. Being an aggressive tumour excision is rarely advised and chemotherapy is the main line of treatment.

References

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