Case report

Pulmonary synovial sarcoma

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ABSTRACT

Primary Synovial sarcoma of the lung is an extremely rare entity. Our patient was healthy prior to presentation and came in with a short course of dyspnea and was found to have a large synovial sarcoma on the lung.

1. Case presentation

We present the case of a 47 y/o male, social smoker (<10 pack years), non-obese with no significant past medical/family history who presented to our office with three months of increasing shortness of breath, left-sided chest pressure, diminished appetite and fifteen-pound weight loss.

Chest X Ray showed a large mass like density in the left upper lobe with mild mediastinal shift and small left pleural effusion (Fig. 1). CT of the chest showed a large heterogeneously enhancing mass in the left hemithorax measuring 16 × 14 × 16 cm with neovascularity in the margin of the mass along with focal areas of calcification with right mediastinal shift and compression atelectasis of the left lung (Fig. 2). A separate 3 × 2 × 2 cm mass was present in the left lung apex. The masses appeared extrapulmonic/pleura based. PET scan showed abnormal FDG uptake with SUV max of 6.6 in the larger mass and a SUV max of 3.4 in the left upper lobe mass.

Using CT guidance, core biopsies were obtained.

Pathology showed spindle cell neoplasm consistent with synovial sarcoma (Fig. 3).

Immunostains showed a monomorphic spindle cell proliferation positive for TLE-1.

Cytokeratin, S-100, TTF-1, desmin CD 117 and CD 34 were negative. H3K27 showed no loss with intact staining. FISH study for SYT gene rearrangement was positive confirming the diagnosis.

Pulmonary function testing showed mild restrictive lung disease, normal volumes and diffusion.

MRI brain was normal.

After a complete staging work up, a diagnosis of monophasic synovial sarcoma involving the left hemithorax, centrally and in the left lung apex, likely pleural based and extrapulmonic was made. Provisional stage was cT3 (unifocal and invading adjacent organ) or cT4 (multifocal), cN0, cM0, grade indeterminate. FISH was positive for SYT gene rearrangement (18q11.2).

The case was presented in the thoracic oncology tumor board and the plan is to do neoadjuvant chemotherapy with epirubicin and ifosfamide followed with thoracotomy and resection.

The patient has tolerated first cycle of chemotherapy well.

2. Discussion

Primary pulmonary diseases can have atypical presentations [1,2]. Lung cancer is the leading cause of cancer related mortality worldwide [3,4]. Primary pulmonary sarcomas account for < 0.1% of all lung cancers and are not associated with cigarette smoking [5] [6]. Primary Synovial sarcomas can have four patterns: monophasic fibrous (spindle-cell), monophasic epithelial, biphasic, and the poorly differentiated monophasic subtype [7]. The available literature on the tumor is sparse and mostly includes case reports. Chest pain and dyspnea are the most commonly described presenting symptoms. Chest imaging usually shows a large well demarcated mass lesion with mediastinal shift/pleural effusion. CT guided or bronchoscopic biopsies are used to get a tissue diagnosis. Immunohistochemistry is used to confirm the diagnosis. The cytogenetic hallmark of synovial sarcoma is the t (X;18)(p11;q11) chromosomal translocation, leading to the rearrangement of the SS18 and one of the SSX genes [8].
surgical resection is often not possible upfront due to the tumor size. Neo adjuvant chemotherapy followed with resection has been described with success.

Overall five year survival is < 50%. Poor prognostic factors include a > 5 cm tumor, higher grade, male sex, older age, neurovascular invasion, and the SYT-SSX1 variant on cytogenetic studies [9].

References

[1] A. Gupta, A.V. Palkar, P. Narwal, Case of chest pain in a young man, BMJ Case Rep. 2018 (2018 Jan 12).
[2] A. Gupta, S. Gulati, Mesalamine induced eosinophilic pneumonia, Respir. Med. Case Rep. 21 (2017 Apr 12) 116–117.
[3] A.V. Palkar, A. Gupta, Y. Greenstein, E. Gottesman, Primary cardiac angiosarcoma: a rare cause of diffuse alveolar hemorrhage, BMJ Case Rep. 2018 (2018 Jun 4).
[4] A. Gupta, A. Palkar, P. Narwal, Papillary lung adenocarcinaoma with psammomatous calcifications, Respir. Med. Case Rep. 25 (2018 Jul 24) 89–90.
[5] R.F. Falkenstern-Ge, M. Kimmich, A. Grabner, H. Horn, G. Friedel, G. Ott, M. Koblihaufl, Primary pulmonary synovial sarcoma: a rare primary pulmonary tumor, Lung 192 (1) (2014 Feb) 211–214.
[6] K. Aydogdu, F. Sahin, G. Fındık, S. Kaya, Pulmonary synovial sarcoma, Asian Cardiovasc. Thorac. Ann. 22 (1) (2014 Jan) 92–94.
[7] S. Okamoto, M. Hisaoka, T. Daì, K. Hatakeyama, T. Iwamasa, H.A. Hashimoto, et al., Primary pulmonary synovial sarcoma: a clinicopathologic, immunohistochemical, and molecular study of 11 cases, Hum. Pathol. 35 (2004) 850–856.
[8] P. Alcârce-Garcìa, S. De ñaz-Palacios, C. Castilìo-Canto, A. Gañàca Pe ñez, J.A. Sa ínez-Gonzàlez Pe ñez, Primary pulmonary biphasic synovial sarcoma: a case report and literature review, Cir. Cir. 80 (1) (2012) 67–71.
[9] M. Trassard, V. Le Doùssal, K. Hacene, Prognostic factors in localized primary synovial sarcoma: a multicenter study of 128 adult patients, J. Clin. Oncol. 19 (2001) 525–534.