CASE REPORTS

ENDOBRONCHIC FOLLICULAR DENDRITIC CELL SARCOMA: AN EXCEPTIONNAL ENDOBRONCHIC TUMOUR

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Abstract

Follicular dendritic cell sarcoma is a very rare entity subject of few studies, we report a case of endobronchial site manifested by a total atelectasis of the right lung managed by initial endobronchic desobstruction and surgical excision with successful results.

Keywords: *sarcoma, endobronchic tumor, lung collapse*

INTRODUCTION

Follicular dendritic cell sarcoma is a very rare thoracic tumor. We report the clinical observation of a young woman in whom bronchial follicular dendritic cell sarcoma was discovered following right lung atelectasis.

OBSERVATION

A 22-year-old woman with no previous history reported a dry cough with progressively worsening dyspnea for 1 month prior to hospital admission. The patient was conscious at the time of examination, with a respiratory rate of 20 cycles per minute and saturation of 95% on room air. Pulmonary examination revealed immobility on the right side of the thorax and a lung collapse syndrome on the right thorax. The lymph nodes were free, and the rest of the clinical examination was unremarkable.

RESULTS

Chest X-ray showed total right pulmonary atelectasis with mediastinal shift (figure 1a). Chest CT confirmed the lung collapse secondary to a suspicious 3 cm tumour obstructing the distal right stem bronchus (figure 1b). Bronchoscopy revealed a round, smooth-walled tumour, hypervascularized, bleeding easily on contact and totally obstructing the bronchial lumen located 3cm from the carina on the right upper lobar bronchus, simulating a carcinoid tumour (figure 1c). The endoscopic appearance of the left bronchial tree was without abnormality, the tumour was excised incompletely because of bleeding and the specimen sent for histological study. The post bronchoscopic course was characterized by a lung expansion . On 18-fluorodeoxyglucose positron emission tomography/computed tomography the remaining endobronchic nodule was isolated and measured about 1.5cm with increased metabolism (SUV=13) without suggestive regrowth (figure 1d).

The pathological result showed architectural tumor proliferation, with tumor cells and an elongated nucleus, fine chromatin and sparse, poorly bounded cytoplasm. Mitoses are rare (1/10 fields). Lymphocytes are present, with no plasma cells or eosinophils. The immunohistochemical study shows partial positive marking of tumor cells by CD23, negative marking for CD21, CD68 , CD3, CD4, CD15, PS100, SOX10, CD34, CD31, CD45, STAT6, EMA, CKAE1/AE3,

chromogranine et synaptophysine, a Ki-67 proliferation index was estimated at 5% of tumor cells (figure 2) compatible with follicular dendritic cell sarcoma. Surgical excision was decided by right upper sleeve lobectomy, the tumor was native from the origin of the upper lobe bronchus. Peroperative pathological control of the margin was free of tumor suggestive of complete removal. In the post operative day1, the chest Xray showed a paramediastinal located effusion without aggravation. Postoperative endoscopy revealed no residual tumor and good bronchial stitches. A 18 FDG-PET was performed 5 weeks after surgery to investigate this remaining effusion and was favorable of post operative sequale. To prevent recurrence and after a multidisciplinary team discussion adjuvant radiotherapy was decided event with complete removal of tumor because of the frequent recurrence described in the literature. Follow-up of the patient 6 months after surgery revealed no recurrence.

DISCUSSION

Follicular dendritic cell sarcoma (FDCS) is a rare low-grade malignancy tumor derived from follicular dendritic cells that serve as accessory cells to the lymphatic system[1-3]. FDCS are part of the four subtypes derived histologically and genetically from stromal or mesenchymal cells, the other 3 subtypes are IDCS (indeterminate dendritic cell sarcoma), FRCT (Fibroblastic Reticular Cell tumor) and DJX (Disseminated Juvenile Xanthogranuloma)[3]. The lymph node sites most often affected are the cervical, axillary and abdominal region; extranodal involvement often concerns liver, spleen,

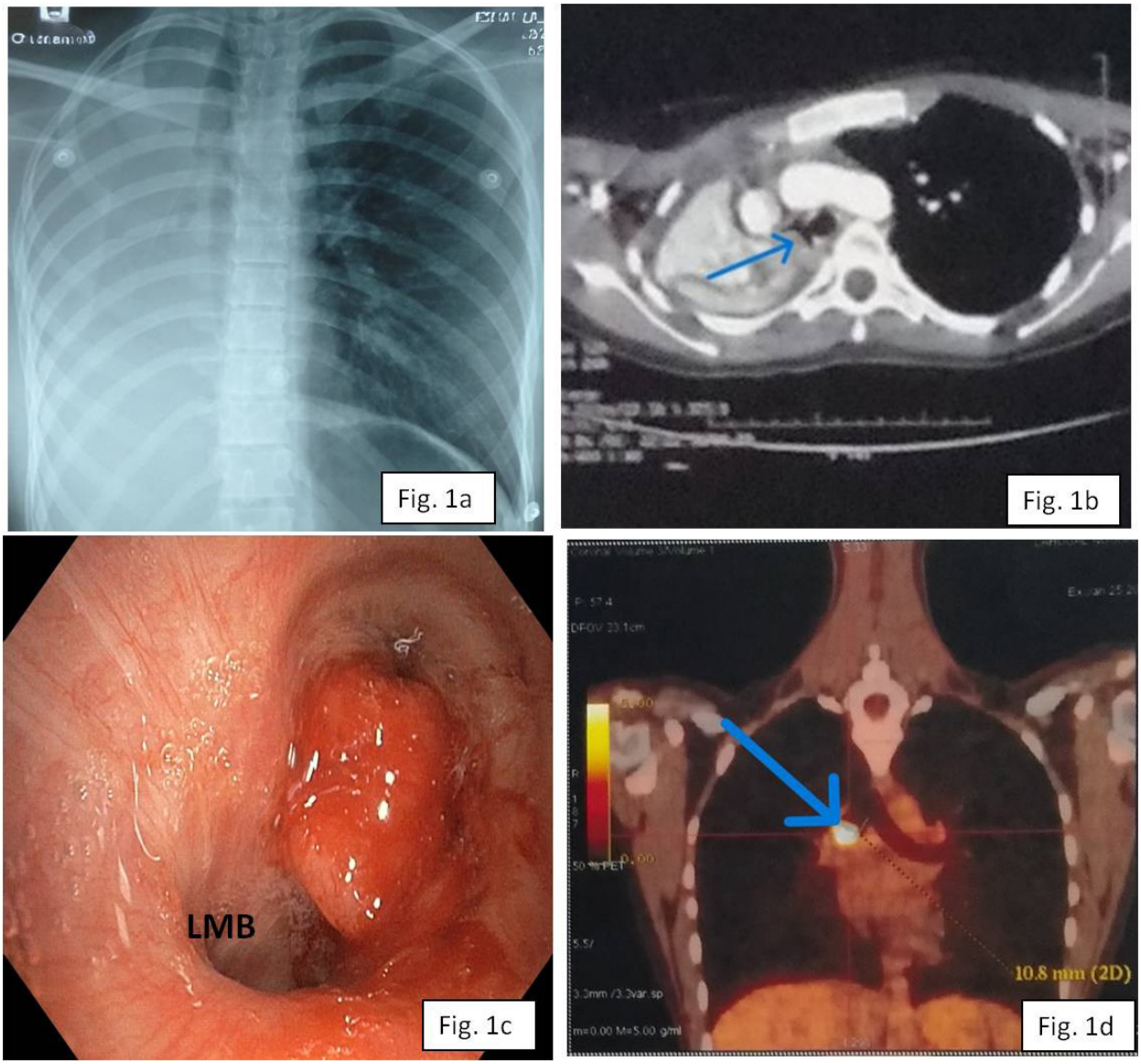

Figure 1

a) initial chest X-Ray before endoscopic excision showing right lung atelectasis;
b) CT scan with total right lung atelectasis with endobronchial tumor (blue arrows);
c) bronchoscopic view showing the tumor lesion in the entrance to the intermediate bronchus;
d) 18 FDG PET bronchial tumor nodule indicated by blue arrows (SUV max = 13).

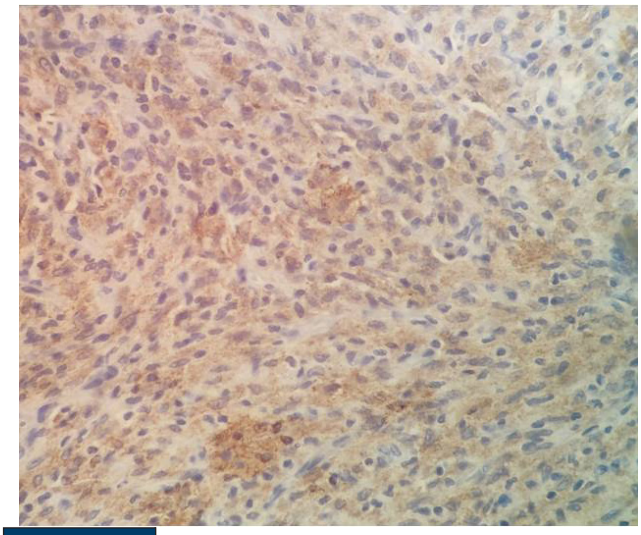

Figure 2 *Pathological view with positive marking of tumor cells by CD23 : magnification x 400.*

gastrointestinal tract, tonsils, mediastinum and lungs[2]. The etiopathogenesis of these sarcomas remains unknown[2,4,5]. FDCS are rarely associated with lymphomas or leukemias[6]. A pseudotumoral inflammatory variant often associated with the Epstein Barr virus exists, especially in women[2-4,7].

The medium age is 49 years with variations from 9 to 90 years, pediatric observations are rare. There is no predilection for sex (sex ratio=1) [4,5]. The clinical presentation is different depending on the organ affected [2,3]. Thoracic involvement was described as a tumor of the chest wall [5] or a large anterior mediastinal mass or posterior mediastinal and para-aortic mass[1]. Bronchial involvement is extremely rare and has been described in a single observation reported by Vinay V and al. in 2022, he revealed a left pulmonary FDCS with endobronchial spread and mediastinal lymphadenopathy in a 34-year-old man, the main manifestation was chest pain for 3 months[3]. The diagnosis can be made by direct biopsy of the tumor lesion either using a needle[5] or by complete surgical excision[3,7].

The tumor cells present immunoreactivity for CD35 in 92.8% of cases, CD21 in 84% of cases, clusterin in 80% of cases and for CD68 in 66% of cases[4,8]. Other studies have also demonstrated positive marking for CD23, CXCL13 and podoplanin; the highest sensitivity concerns CD-21, CXCL13 and clusterine in 80% of follicular dendritic cell sarcoma[9]. Differential diagnoses includs large cell lymphoma, mesenchymal tumors, solitary fibrous tumors and leiomyomas[2,5].

Surgery represents the main option for radical treatment alone or combined with radiotherapy and chemotherapy but with uncertain role[2,3]. The rare observations reporting thoracic involvement note the use of pneumonectomy in cases of parenchymal involvement, especially in cases of large tumors[3], surgery offers a better prognosis for the disease. The use of neo-adjuvant radiotherapy was made to reduce the resection margins, adjuvant radiotherapy was prescribed in cases of narrow resection margins[1], the cumulative dose can variate from 30 to 63 Gy administered on 30 to 35 fractions. Chemotherapy and targeted therapies may be used at the metastatic stage[8]. The postoperative prognosis mainly depends on the extent of the disease, surgical excision and histopathological characteristics[2]. The median survival is estimated at 2,9 years for all stages FDCS combined. The 2-year survival for a localized tumor is estimated at 82.4%, 80% for a locally advanced stage and 42.8% for metastatic stage[4].

CONCLUSION

Follicular dendritic cell sarcoma manifested by an endobronchial tumor with total atelectasis of the right lung remains a very rare entity. This observation highlights another clinical and management feature of the disease.

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