Int J Clin Exp Pathol 2021;14(11):1102-1105
www.ijcep.com /ISSN:1936-2625/IJCEP0039360

Case Report

Pleuro-pulmonary blastoma in a 45 year-old woman after Hodgkin's lymphoma: a case report

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Received September 3, 2016; Accepted October 24, 2016; Epub November 15, 2021; Published November 30, 2021

Abstract: Pleuro-pulmonary blastoma is a rare and aggressive intrathoracic mesenchymal tumor occurring in childhood. We report the exceptional case of a forty-five year-old woman, presenting with respiratory distress. She had previously been treated with chemotherapy and radiotherapy for Hodgkin lymphoma. Chest X rays and computed tomography scan showed a left lung lower lobe mass and pleural effusion. Biopsy cores revealed a pleura-pulmonary blastoma tumor with mixed components. Neoadjuvant chemotherapy was decided in multidisciplinary round. Because of the rarity of these tumors, no consensus for the treatment exists.

Keywords: Pleuro-pulmonary blastoma, Hodgkin's lymphoma, second-cancer, radiotherapy, lungs

Introduction

Pleuro-pulmonary blastoma is a rare and highly aggressive intra-thoracic tumor accounting for 15% of all primary pediatric pulmonary tumors. It affects generally infants under four years [1].

We present the case of a Caucasian woman aged of 45 years, presenting with a pleuro-pulmonary blastoma, revealed by thoracic pain, after she had received chemotherapy and radiotherapy for Hodgkin's lymphoma fifteen years ago.

Case presentation

A 45-year-old Caucasian female presented to emergency with sudden thoracic pain and breathing difficulties.

She had been suffered from dyspnea for several weeks.

She had smoked one packet a day, for 20 years. She had no surgical or cancer familial past.

She had been treated fifteen years ago, at age of 30, for grade III, disseminated Hodgkin's lymphoma. She had received chemotherapy with ABVD regimen (Adriamycin; Bleomycin; Vinblastine and Dacarbazine) and mantle radiotherapy.

At clinical examination, the patient presented with retrosternal pain and left basis pulmonary abolition murmur.

Cardiac origins of the dyspnea were excluded.

Chest X rays revealed a left pleural effusion with passive collapses and atelectasis, and a left lower lobe density. **Figure 1**.

Computed tomography scan showed the pleural effusion, but did not notice any mass. There was no sign of pulmonary embolism. **Figure 2**.

The patient underwent percutaneous pleural. Two litres of hematic fluid were evacuated. No malignant cell were found at microscopic staining.

Positron emission tomography scan was performed and showed a high hypermetabolic (SUV max 20.6) 46 mm mass originating from the lingula and the left lower lobe, and a infra-centimetric hypermetabolic (SUV 4.5) coeliac node. The pleural effusion was not hypermetabolic. The tumor was abutting but did not involved pleura. **Figures 3** and **4**.

Brain magnetic resonance imaging didn't found any metastasis.

Transthoracic ultra-sound guided biopsy was performed without any complications.

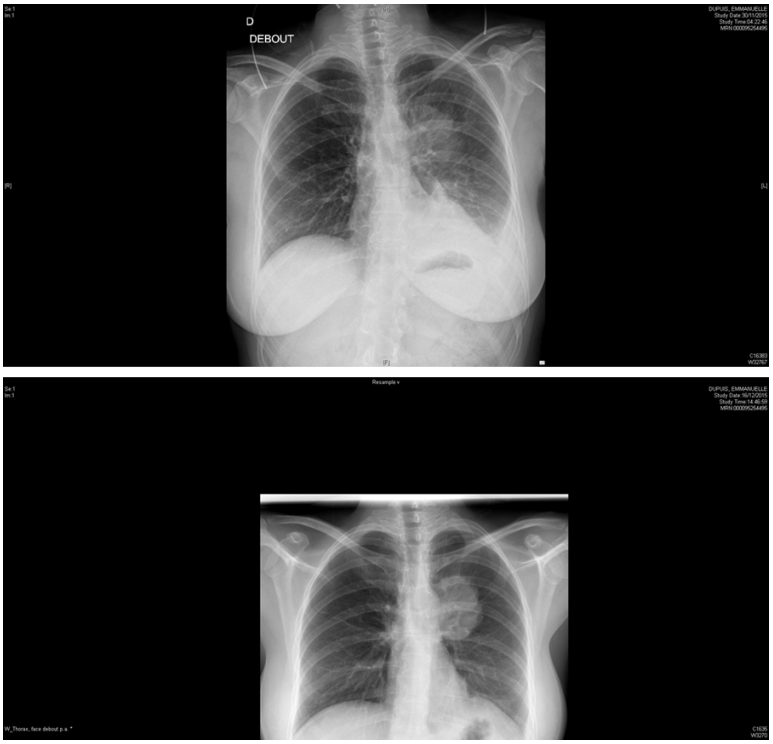

Figure 1. Chest radiography at presentation: left minime pleural effusion and left lower lobe density.

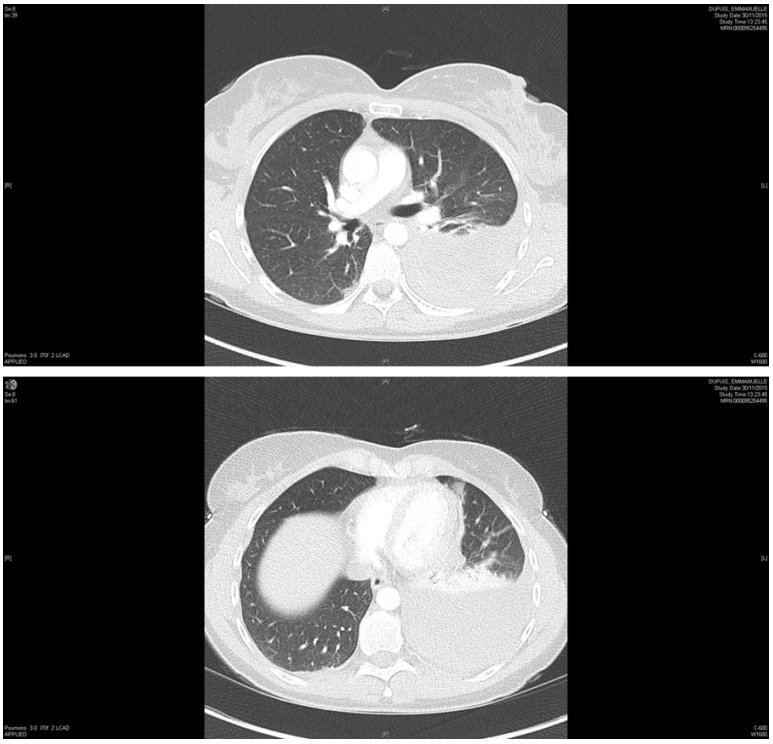

Figure 2. Computed-Tomography scan showing left pleura effusion.

Biopsy cores were obtained. They were multiple fragments of pulmonary tumor, characterized by an heterogeneous polymorphic aspect, made of several cells components. Glandular structures with basal nucleus, surrounded by fusocellular stroma composed by entangled cells and showed many scattered mitoses. That proliferation showed "microrosette" architecture.

It was showed an epidermoid component closely mixed up in fusocellular stroma like morula. No necrosis or inflammation area was found.

Immunohistochemically, the glandular component was positive for ACE, TTF1, CK7, CD99, Chromogranin, CKAE1/AE3, EMA and Synaptophysin.

The epidermoid morula were positive for P40 and CK5/6.

The mesenchal component was positive for Desmin and Myogenin. AML was negative. Glypican3, PLAP and alpha-foetoprotein were focally positive. Beta catenin was strongly positive in the cytoplasm of all the cells.

Estrogen and progesterone receptors were negative, PAX8, CD117, PS100, MDM2 were all negative.

Ki-67 was very high, at 60%.

On that basis, the diagnosis of malignant pleuro-pulmonary blastoma with double glandular and mesenchymal component like heterologous component such as rhabdomyosarcoma, was made and the material was sent to Nancy for confirmation. There it was confirmed to be pleuro-pulmonary blastoma with massive rhabdomyoid component.

Treatment options were discussed in multidisciplinary round.

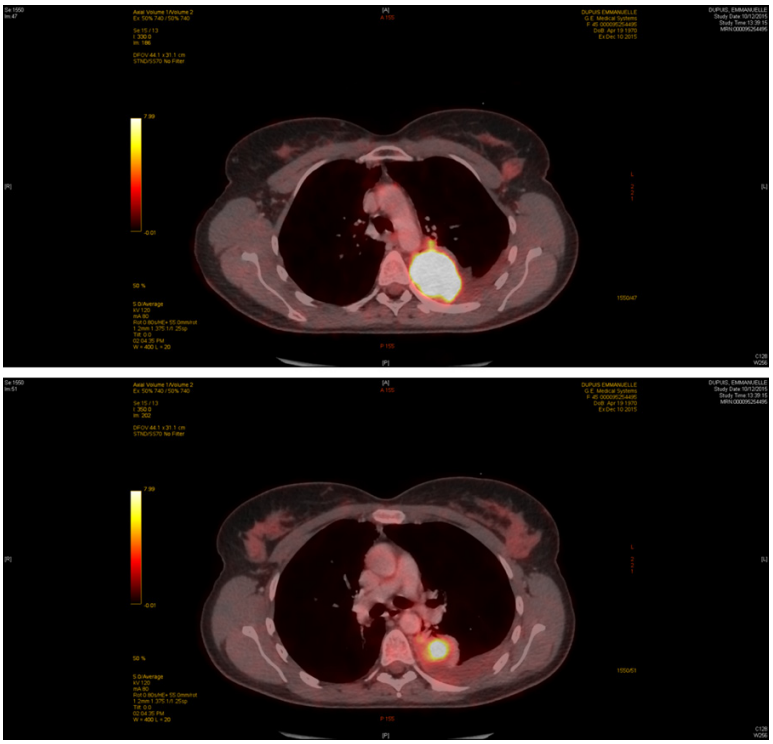

Figure 3. Positron Emission Tomography: high hypermetabolic (SUV max 20.6) 46 mm mass originating from the lingula and the left lower lobe.

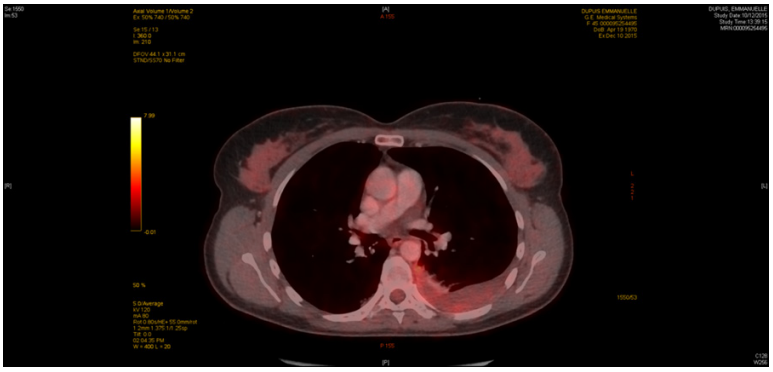

Figure 4. No hypermetabolism of the pleural effusion.

It was decided to realize neoadjuvant chemotherapy according IVADo regimen (Ifosfamide+ Vincristin+Actinomycin D and Doxorubicin) followed by surgery, if feasible.

Discussion

Pleuro-pulmonary blastoma is a rare intrathoracic tumor accounting for 15% of all pediatric pulmonary tumors. The age of presentation is usually young, under 4 years. Very few cases have been reported in patients over 10 years and only one case has been reported at 36 years. To best of our knowledge, less than 100 cases have been reported in the literature.

We have described the second case in adult patient and the first case, to best of our knowledge, occurring after Hodgkin's lymphoma.

It was first reported in a study of 11 patients by Manivel et al. in 1988 [2], but Spencer first used the term in 1961, suggesting that those tumors arose from mesodermal blastoma, because similarities with nephroblastoma.

These tumors are known to be aggressive with tendency to spread to the brain.

Three types of pleura-pulmonary blastoma exist, based on morphology.

Type I, accounting for 15 to 20% of all pleura-pulmonary blastoma have most favorable prognosis, while types II and III have poorest outcomes.

Type I tumors are purely cystic, peripherally located without chest wall invasion. Microscopically, the cysts are round or ovale and lined by cuboidal or columnar ciliated epithelium; the subepithelium may contain layer of primitive sarcoma with scattered rhabdomyoblasts.

Type Ir (type I-regressed) tumors are cystic containing few spindle shaped cells with foci of dystrophic calcification without subepithelial malignant cell condensation. Only 8% of such tumors progress to type II or III.

Type II tumors are partly solid and cystic sharing features of both type I and II tumors. On microscopic examination, the cysts are same as type I. However the solid part is identical to

type III tumors. The tumors have plaque-like or nodular proliferation in the cyst wall composed of blastema-like cells or malignant cells with or without rhabdomyoblasts in the subepithelium.

Type III tumors are entirely solid tumors usually presenting as large masses well-circumscribed that can be friable due to hemorrhages and necrosis. There are blastematous foci and frequent mitoses. The sarcomatous component is composed of spindle cell proliferation. Rhabdomyoblasts occur singly or in clusters.

Several cytogenetic changes such as trisomy 2, 8, and P53 mutation have been reported that the last one had fatal outcome [3].

The median age of presentation depends on the stage of the tumor. Tumors affect male and female patients without gender predilection and occur more commonly on the right side.

Clinically patients present with chest or abdominal pain, fever, dyspnea, cough, hemoptysis or general symptoms.

Immunohistochemical staining is not mandatory for diagnosis but can help.

Brain metastases are common, that's why it is mandatory to follow up these patients by brain imaging studies [4].

Recommended treatments for type I consist of surgical excision and adjuvant chemotherapy. The type Ir are treated only by surgery without chemotherapy. Type II and Type III, should receive 2 or 4 courses of neoadjuvant chemotherapy to reduce tumor size, that is obtained in more than 90%, before being resected. The recommended chemotherapeutic agents are Ifosfamide, Vincristin, Actinomycin D and Doxorubicin (IVADo).

Recurence of disease are treated with high dose chemotherapy regimen and autologous stem cell rescue.

Radiation therapy is reserved for patients with nonresectable disease or for residual tumors after chemotherapy [5].

The prognosis depends largely on the staging at the time of diagnosis and the grading sarcomatous elements. The 5-year survival is probably less than 50% [6].

Our case is exceptional because the older age of presentation disease.

To the best of our knowledge it is the first case reported after treatment of Hodgkin lymphoma. The link between the tumor and radiation therapy and/or chemotherapy is not known.

Disclosure of conflict of interest

None.

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