

Desmoplastic small round cell tumor of pleura: Case report and review of literature

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Abstract

Desmoplastic small round cell tumor (DSRCT) is a rare malignant neoplasm of young adults. We report a case of 50-year-old woman with metastatic pleuro-pulmonary desmoplastic small round cell tumour who underwent palliative chemotherapy with an initial clinical improvement and radiologic stability of the disease. Nowadays, there is still no clear consensus regarding optimal treatment of desmoplastic small round cell tumours. Most papers reinforce the importance of multimodality treatment consisting of high-dose chemotherapy, surgical resection and radiotherapy for localized tumours.

Introduction

Desmoplastic round cell tumour (DRCT) is a distinct clinico-pathological entity that has been described recently (1,2). This aggressive malignant neoplasm tends to affect young adults and occurs predominantly in the abdomen. DSRCT in the lung is extremely rare. In this article, we report a pleuro-pulmonary DSRCT with its histochemical, immunohistochemical and cytogenetic features diagnostic and we discuss its treatment.

Case report

A 50 years old woman who had cough and dyspnea progressively worsening two months before consultation. There was no history of smoking or exposure to other carcinogens. Clinical examination showed a syndrome of pleural effusion. Chest Radiography revealed multiple nodular pulmonary lesions, associated with a partial lung collapse, calcifications and a left pleural effusion (Fig. 1). The patient had thoracoscopy with nodular biopsy. Histologically, nests of small round cells surrounded by intense desmoplastic reaction were seen in pleura extending along the septa into the lung parenchyma. Immunohistochemical staining revealed coexpression of cytokeratins, vimentin, desmin, CD 99 and neuron-specific enolase in the tumor. Cytogenetic study objectived translocation t(11;22) confirming the diagnosis of pleuropulmonary desmoplastic small round cell tumour. The patient was treated with Adriamycin (60 mg/m² day 1), Ifosfamide (3000 mg / m²: D1 to D3) every three weeks with mesna and GCSF. Evaluation after 6 cycles showed significant clinical improvement and a

radiologic stability of the disease (figure 2). Then the patient was treated with cyclophosphamide administered orally in maintenance.

Comments

Desmoplastic small round cell tumor is a polyphenotypic mesenchymal neoplasm associated with a prominent fibrous stroma (1- 2). It is a highly aggressive tumor which was described in 1989 by Gerald and Rosai (3) and Ordonez et al (4). It typically affects young with male predilection and peak incidence in the second decade (2). DSRCT most often arise in peritoneum and mesothelial surfaces and predominant in the abdomen. Extra abdominal DSRCT is rare and includes brain, pleura, lung, nasal cavity, salivary gland and soft tissue and bone (5). To date, only two cases of pulmonary DSRCT and six cases of pleura were reported in English literature (6-13). The patients with pleural and pulmonary DSRCT usually presented with cough chest pain and pleural effusion. There are no reliable and specific biomarkers for its diagnosis which is established with correlation of radiological, histological, immunohistochemical and molecular features: CT scan is the most commonly radiologic technique used to approach a diagnosis of the DSRCT of lungs and pleura. At histological analysis, the tumour manifests as islands of small blue cells surrounded by fibrous stroma. It should be considered in the differential diagnosis of round cell tumours such as Ewing's sarcoma, neuroblastoma, Wilm's tumour, rhabdomyosarcoma and primitive neuroectodermal tumour (14).

On immunohistochemical studies, DRCT are immunoreactive with antibodies to desmin, vimentine, keratin, epithelial membrane antigen, and neuronspecific enolase. The histological and immunohistochemical features in our case are similar to those described in the literature. Cytogenetically, DRCT is associated with a specific chromosomal abnormality, t (11;22) (p13: q12)(14-16). The breakpoints involve the EWS gene on 22q12 and Wilms gene (WT1) on 11q13 (6). Recently, a novel EWS/WT1 gene fusion product, EWS-WT1 5/10 was reported. Recently, WT1, a polyclonal antibody against the amino terminus of the WT1 protein, has shown positive staining in more than 95% of cases, as was seen in our case (7).

The prognosis of DRCT is very poor. : the median overall survival of patients reported from summarized results on prognosis is 17 months (17). These tumours occur an aggressive multimodality therapy consisting of high-dose chemotherapy, surgical resection and radiotherapy. Surgery is often not feasible

because of the extensive involvement (18). There are only a few reports on the role of chemotherapy in these Patients. A complete response after a chemotherapy regimen consisting of vincristine, doxorubicine, cyclophosphamide, ifosfamide and etoposide in 10 patients with DSRCT was observed (19). However, high dose chemotherapy with stem cell transplantation didn't demonstrate superiority to conventional chemotherapy in term of overall survival in patients with DSRCT (20, 21). Radiotherapy completing neoadjuvant chemotherapy and surgery had improved the median overall survival to 32 months (22).

Our patient was treated with combination chemotherapy consisting of doxorubicine and ifosfamide as first line palliative chemotherapy and subsequently treated with cyclophosphamide administered orally in maintenance.

Figures



Fig. 1: CT of thorax revealing nodular lesions in right lung associated with a left pleural effusion (Before chemotherapy)



Fig. 2: CT of thorax revealing stability of nodular lesions and pleural effusion after 6 cycles of chemotherapy

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