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(CASE REPORT) (RESEARCH ARTICLE)

Intrapulmonary desmoplastic mesothelioma in a young adult – Case report

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Abstract

Introduction: Desmoplastic mesothelioma typically represents an extremely rare histological subtype of sarcomatoid malignant mesothelioma that usually presents with a diffuse pattern of growth. A marked increase in mesothelioma is predicted in developing countries, where asbestos usage is increasing exponentially. Only a few case series have been reported in the Indian literature where mesotheliomas have been analysed on routine histology and IHC.

Case Presentation: We are reporting a case of a 28 years old man came with complaints of gradually progressive central non radiating chest pain for 3months with unremarkable physical findings. CT thorax revealed well defined homogenous slightly lobulated soft tissue attenuating mass lesion in anterior epicardial fat pad, biopsy reported benign spindle cell tumour possibly neural origin.

Interventions: Patient underwent thoracotomy and tumour excision. Post-operative histopathological examination showed desmoplastic mesothelioma. The final diagnosis of intrapulmonary desmoplastic mesothelioma was confirmed by pathological and immunohistochemical examination. The patient underwent 6 cycles of PEM/CARBO adjuvant chemotherapy followed by adjuvant radiation of 50.4 Gy by VMAT Technique.

Conclusion: Desmoplastic mesothelioma is a rare pathology and rarely seen in young patients. Desmoplastic mesothelioma continues to have a poor prognosis due to its highly malignant, aggressive, and refractory nature to local treatment which necessitates the early intervention to improve the survival.

Keywords: Desmoplastic; Mesothelioma; Chemotherapy; Radiotherapy

1. Introduction

Desmoplastic mesothelioma typically represents an extremely rare histological subtype of sarcomatoid malignant mesothelioma that usually presents with a diffuse pattern of growth [1]. A marked increase in mesothelioma is predicted in developing countries where asbestos usage is increasing exponentially. Asbestos exposure, often as an occupational hazard, has been clearly linked to the occurrence of malignant pleural mesothelioma (MPM [2]. The case of localized intrapulmonary desmoplastic mesothelioma is especially rare. This type of tumour lies in between epithelioid and sarcomatous subtypes of mesothelioma and may mimic fibrous pleuritis. Only a few case series have been reported in the Indian literature where mesotheliomas have been analysed on routine histology and IHC.

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2. Case report

A case of a 28 years old man came with complaints of gradually progressive central non radiating chest pain for 3 months with unremarkable physical findings. CT thorax revealed well defined homogenous slightly lobulated soft tissue attenuating mass lesion in anterior epicardial fat pad. CT guided Biopsy reported benign spindle cell tumour possibly neural origin. Patient underwent thoracotomy and tumor excision under general anaesthesia. Post-operative histopathological examination revealed findings suggestive of desmoplastic mesothelioma. The final diagnosis of intrapulmonary desmoplastic mesothelioma was confirmed by pathological and immunohistochemical examination. The patient underwent 6 cycles of PEM/CARBO adjuvant chemotherapy followed by adjuvant radiation of 50.4Gy/28 fractions with 1.8 Gy /fraction. Histopathology and immunochemistry shown in figure 1

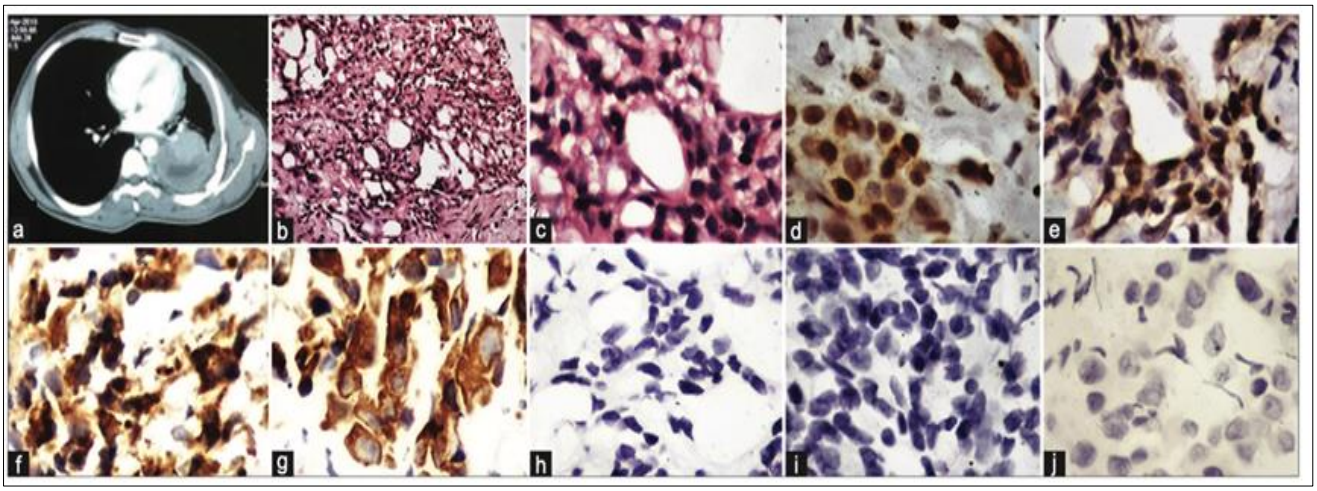

Figure 1 Epithelioid mesothelioma. (a) CECT thorax shows loss of ling volume on the left side with thickwned nodular pleura. (b andc) polygonal cells infiltrating fibroadipose tissue (Hand E; ×100 and 400). ((d-g) WT1, calretinin, D2-40, and CK5/6 showing positivity in the tumor cells (polyHRP; × 400 (h-j) tumor cells negative for ttf1, napsin, and CEA (polyHRP; ×400)

2.1. CECT Report of Thorax

CT thorax revealed well defined homogenous slightly lobulated soft tissue attenuating mass lesion in anterior epicardial fat pad (blue arrow) and repeat CT thorax showed regression of tumour following treatment (red arrow) figure 2

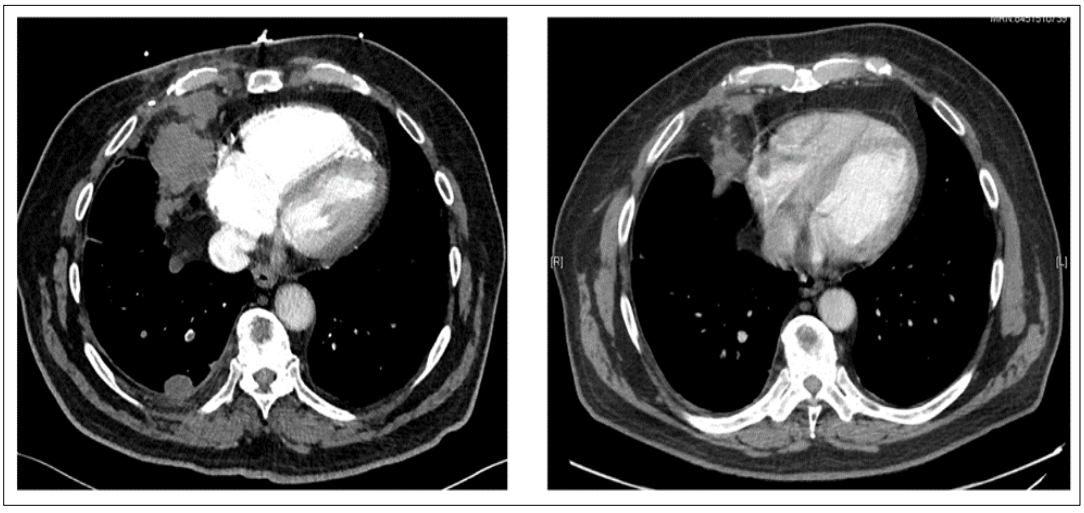

Figure 2 CECT of thorax pre-treatment (blue arrow) and post treatment (red arrow)

2.2. Treatment Fields with Vmat Technique

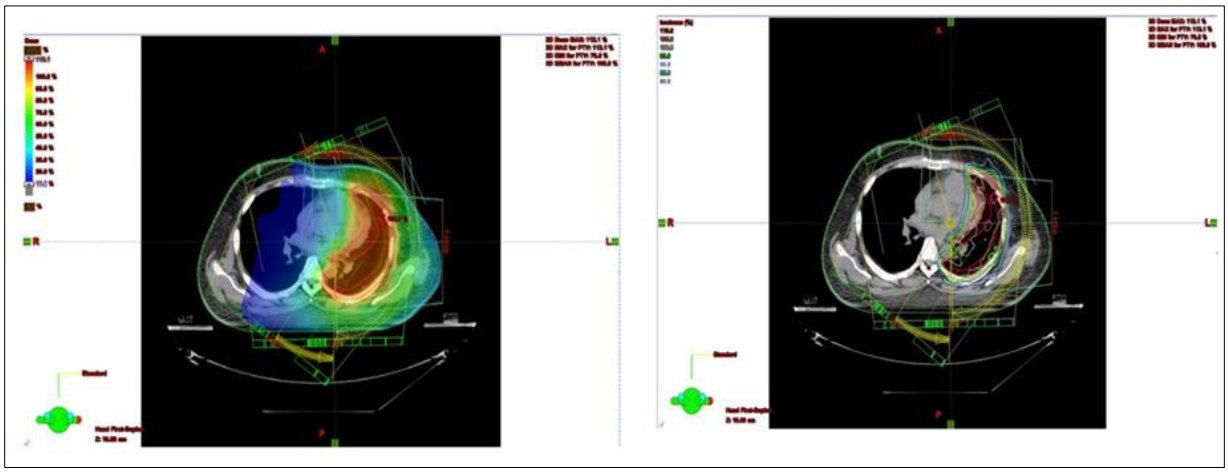

Figure 3 Patient was treated with conformal VMAT technique with a dose of 54 GY with 1.8 Gy/fraction 5 day per week for a period of of 6 weeks treatment field included 3 arcs

3. Discussion

Desmoplastic mesothelioma is a rare variant of malignant mesothelioma with a storiform collagen pattern, collagen necrosis, bland acellular collagen and focal cytological features of malignancy. MMs are histologically heterogeneous tumors predominantly seen in males with a peak age incidence in the fifth to sixth decade. The most common etiology in cases of pleural and peritoneal mesotheliomas is linked to asbestos exposure with a long period of latency ranging from 30 to 40 years. It arising most frequently in the pleura, peritoneum or pericardium and less frequently in organs such as lung, spleen, pancreas and liver. In fact, few cases of intrapulmonary malignant mesothelioma have been described presenting with bilateral miliary pulmonary nodules and diffuse bilateral opacities or characterized by diffuse intrapulmonary growth [3].

Sonja Klebe, et al [4] Reviewed 326 sarcomatoid malignant mesotheliomas with 70 cases (21%) classified as DMM and without a case arising in lung and found it occurred more commonly in men. It is essential to differentiate intrapulmonary DMM from other pulmonary tumors, such as sarcomatoid carcinoma, because of important differences in treatment and prognosis. Pathological diagnosis is the most reliable diagnostic method. And immunohistochemical staining can aid in making the diagnosis by providing additional characteristic histological information, in that DMM is positive for calretinin and D2- 40, but negative for carcino-embryonic antigen [5].

Treatments for DMM aim to reduce symptoms, prolong survival and improve quality of life without taking aggressive action. The most common therapy options are chemotherapy, radiation and surgery, which shrink tumours and kill cancerous cells. Currently, DMM without definitive treatment is generally managed according to mesothelioma guidelines. NCCN guidelines recommend that patient with malignant mesothelioma should receive chemotherapy. Induction or the adjuvant therapy approach for malignant mesothelioma has not been tested in a randomised study and the relative contribution of chemotherapy and/or radiotherapy given before or after radical debulking surgery is largely unknown. On the other hand, there are observations that platinum plus pemetrexed chemotherapy is occasionally able to induce a complete pathologic response and that the addition of chemotherapy and/or radiotherapy to radical surgery is associated with more favourable outcomes [6].

Radiotherapy is widely used in the treatment of patients with malignant mesothelioma. This patient was treated with conformal VMAT technique with a dose of 50.4Gy/28 fractions with 1.8 Gy /fraction 5 days per week for a period of 5.5 weeks. Treatment field included 3 arcs of clockwise 340- 180 degree, counter-clockwise 0f 180-340 degree and 215-181 degree arch to reduce the low dose exit dose spillage to the contralateral side. It is used for palliation of symptoms such as pain, for port-site prophylaxis and is considered an integral part of multimodality therapy for early stage disease where it may prevent local relapses after surgical resection.

4. Conclusion

Desmoplastic mesothelioma is a rare pathology and rarely seen in young patients. Desmoplastic mesothelioma continues to have a poor prognosis due to its highly malignant, aggressive, and refractory nature to local treatment which necessitates the early intervention to improve the survival.

Compliance with ethical standards

Acknowledgments

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Disclosure of conflict of interest

None

Statement of informed consent

Informed consent was obtained from patient included in the study.

References

[1] Galateau-Salle F, Churg A, Roggli V, Travis WD, World Health Organization Committee for Tumors of the Pleura. The 2015 World Health Organization Classification of Tumors of the Pleura: Advances since the 2004 Classifi cation. J Thorac Oncol. 2016; 11: 142-154.

[2] Bianchi C, Bianchi T. Global mesothelioma epidemic: trend and features. Indian J Occup Environ Med. 2014; 18(2): 82–8.

[3] Hida T, Hamasaki M, Matsumoto S, Abe S, Takakura K, et al. Diffuse intrapulmonary malignant mesothelioma presenting with miliary pulmonary nodules: A case report. Pathol Int. 2015; 65: 318-323.

[4] Klebe S, Brownlee NA, Mahar A, Burchette JL, Sporn TA, et al. Sarcomatoid mesothelioma: a clinical-pathologic correlation of 326 cases. Mod Pathol. 2010; 23: 470-479.

[5] Liu M, Wu K, Tan Y. Localized intrapulmonary desmoplastic mesothelioma: A case report. J Radiol Oncol. 2019; 3: 014-016.

[6] Yan TD, Cao CQ, Boyer M, et al. improving survival results after surgical management of malignant pleural mesothelioma: an Australian institution experience. Ann Thorac Cardiovasc Surg. 2011; 17: 243-9.