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Primary Pulmonary Ewing Sarcoma: The Role of Adjuvant Radiotherapy in a Rare Thoracic Malignancy - A Case Report

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Abstract: ***Background: Primary pulmonary Ewing sarcoma (PPES) is exceedingly rare and treatment is extrapolated from skeletal Ewing sarcoma protocols. Radiotherapy is an important component of local control in selected patients with poor pathological response or residual disease after chemotherapy and surgery. [1-5] Case: A 23-year-old man presented with chest pain and dyspnea for 3 months. Imaging showed an 8×6×3 cm right lower lobe mass. After VAC/VIE chemotherapy and right lower lobectomy, histopathology demonstrated approximately 80% viable residual tumor. Adjuvant radiotherapy (45 Gy/25 fractions) with concurrent vincristine followed by chemotherapy was delivered. The patient remains disease free on follow-up. Conclusion: Postoperative radiotherapy may improve local control in patients with poor pathological response. [1,5]***

Keywords: Primary pulmonary Ewing sarcoma, Postoperative radiotherapy, Pathological response, Residual tumor, Local disease control

1. Introduction

Ewing sarcoma is a highly aggressive malignant small round cell tumor predominantly affecting children and young adults. [1,2] Primary pulmonary Ewing sarcoma accounts for less than 1% of reported cases. [1,3] Owing to its rarity, management is extrapolated from conventional Ewing sarcoma protocols. [1,5] Radiotherapy is a key component of local treatment because of the radiosensitive nature of Ewing sarcoma and is recommended for unresectable disease, positive or close margins, poor pathological response, or residual microscopic disease. [1,5] Modern conformal techniques such as 3D-CRT and IMRT improve target coverage while minimizing dose to the lungs, heart, esophagus and spinal cord. [1,5]

2. Case Presentation

A 23-year-old male presented with right-sided chest pain and exertional dyspnea for three months. Chest X-ray showed a well-defined homogeneous opacity in the right lower lung zone. CT thorax demonstrated an 8×6×3 cm right lower lobe mass. CT-guided biopsy with immunohistochemistry (CD99 and FLI-1 positive) confirmed primary pulmonary Ewing sarcoma. After multidisciplinary discussion, neoadjuvant VAC/VIE chemotherapy was administered followed by right lower lobectomy. Histopathology showed approximately 80% viable residual tumor with negative margins. Considering the poor pathological response, adjuvant external beam radiotherapy was delivered to the postoperative tumor bed to 45 Gy in 25 fractions using conformal radiotherapy with concurrent weekly vincristine. Organs at risk including bilateral lungs, heart, esophagus and spinal cord were contoured and respected during planning. Treatment was completed without interruption and followed by additional chemotherapy. The patient remains on regular follow-up without evidence of recurrence.

3. Discussion

The major learning point in this case is the role of postoperative radiotherapy. Although complete resection was achieved, extensive residual viable tumor after induction chemotherapy indicated poor pathological response and a higher risk of local recurrence. Because Ewing sarcoma is radiosensitive, adjuvant radiotherapy was used to eradicate microscopic residual disease and improve local control. Modern conformal planning allows safe treatment while reducing dose to surrounding thoracic organs. This case supports the value of integrating radiotherapy into multidisciplinary management of this rare thoracic malignancy. [1,5]

4. Conclusion

Primary pulmonary Ewing sarcoma requires multidisciplinary treatment. In patients with poor pathological response after neoadjuvant chemotherapy, postoperative radiotherapy should be considered to optimize local control even after complete surgical resection. Reporting such cases helps strengthen the evidence for radiotherapy in this rare disease. [1,5]

References

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