Role of Radiotherapy in Conjunction with Chemotherapy in Giant Pleuropulmonary Blastoma

Pleuropulmonary blastoma is an exceedingly rare childhood malignancy constituting less than 1% of all the pediatric tumors(1). The cell of origin is believed to be the embryonic rests of fetal lung tissue with hamartomatous and sarcomatous elements(2). Despite the introduction of multimodal therapy, it still carries a poor prognosis. Surgery is considered as the primary modality of treatment in pulmonary blastomas (PB). Other treatment modalities like radiotherapy and chemotherapy have been tried but with no significant improvement in the outcome. Here we describe one case of a giant pleuropulmonary blastoma who was treated by multimodality therapy.

A thirteen year old male presented to us with pain and lump right chest wall for two years duration. The lump was insidiously growing and was initially suspected to be tuberculosis by a physician and antitubercular treatment was started. Despite the above treatment the lump started to grow rapidly in the last three months. At the time of presentation, the patient looked uncomfortable with scoliotic posture. He was cachexic with moderate degree of pallor. A large mass measuring 18x15 cm was visible on the back (Fig. 1). This massive mass seemed to be coming from within the thoracic cage, had a smoothening glistening surface, was firm in consistency, non pulsatile and was fixed both to skin and the underlying muscles and rib cage. There was no thrill or bruit over the mass. On auscultation, there was marked diminution of breath sounds in the right mammary and inframammary regions. The rest of the systemic examination was within normal limits. His routine biochemical parameters and blood profile should be to ensure honest selection of the right candidate as a "teacher".

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were within normal limits. X-Ray chest revealed a huge soft tissue mass in the right middle and lower zone with an extension to the adjacent chest wall. There was no mediastinal shift or any other evidence of lymphadenopathy. CECT scan of the chest showed a similar mass extending outside the confines of chest wall and invading the diaphragm and liver on the same side. Fine needle aspiration cytology revealed pleuropulmonary blastoma. In view of the inoperable nature of lesion and keeping the patient's age in mind, a combined protocol of chemotherapy and radiotherapy was decided upon. Combination chemotherapy, i.e., vincristine (1.4 mg/m²), actinomycin-D (15 µg/kg), cyclophosphamide (500 mg/m²) and adriamycin (30 mg/m²) were given in two courses at 3 weekly interval. Subsequently, local radiotherapy to the involved hemithorax was given. A total dose of 30 Gy in 10 fractions was delivered by parallel opposed portals by a telecobalt unit. There was minimal response immediately following the treatment. The disease however, started to progress 2 months after therapy. At present the patient is alive but symptomatic with progressive disease at the end of 6 months from the date of diagnosis.

Pulmonary blastoma was described as early as in 1945 as one of the rarest malignant lung tumors(2). Since than about 40 cases have been reported in the literature. Most of the literature focuses on pathology and classification, but there is a lack of comparable data for treatment evaluation. It is postulated that the tumor originates from mesenchymal pulmonary blastoma, analogous to nephroblastoma. The common histological feature is the presence of primitive cells with blastomatous qualities separated by a stroma which usually has a sarcomatous element. The presence of mesothelial element makes it pleuropulmonary blastoma(3).

Although surgery remains the mainstay of treatment, combined modality approach has been introduced but the results so far continue to remain dismal. One of the major reasons for failure is late presentation, which more often than not leads to incomplete surgical resection. This is due to early involvement of the parietes including adherence of the tumor tissue to the mesothelium and also due to early extension into the mediastinum(4). The role of chemotherapy is still experimental producing variable results(5). Moreover the number of such cases in literature are few. Local radiotherapy has been tried without any better outcome(3). Recently, combination chemotherapy has been tried in conjunction with radiotherapy in an inoperable patient like ours(6). In this case, 75% tumor regres-
sion was observed but the patient died of neutropenic septicemia.

Since this was a huge right lung mass invading the chest wall, diaphragm and liver, surgery was not possible. Radiotherapy in conjunction with chemotherapy was tried. Only minimal response was obtained initially, following which there was rapid progression of the disease. This further indicated that size of the tumor is the most important prognostic factor and it is believed that a size of over 5 cm holds a grave prognosis.

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The recent article on the subject was interesting and informative(I). The outcome must have been influenced to a great extent by the package of services which included mothers' education/awareness on child survival through clinics and outreach activities apart from providing health care and nutrition supplements.

It would have been interesting to know the nutritional status of the cohort (grades and percentage of malnutrition) at the end of the study. This would have shown the morbidity load and the quality of health achieved with the given intervention. It was also important to know the female literacy rate during 1967 and at the time of undertaking the study in 1988-89.

It is now realized more and more that mere distribution of food supplement and providing health care facilities are not enough to achieve our national goal of better health and nutrition for women and children. It is also important to provide inputs for improving education/awareness of the community regarding better environmental hygiene and sanitation and developmental stimulation (social and psycho-