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Sarcomatoid Thymic Carcinoma: A Case Report

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PURPOSE

To evaluate the response of Chemo-radiation in an inoperable sarcomatoid thymic carcinoma.

BACKGROUND

Approximately two-thirds of all primary mediastinal tumours are benign. The most common tumor of the anterior mediastinum is thymoma. Thymic carcinomas are rare epithelial malignancies that have a more aggressive clinical course than thymomas. Thymic carcinomas comprise approximately 15% of thymic tumours. Thymic carcinoma frequently involves the pleura and locoregional lymph nodes. Distant metastases to the lungs, liver, brain, and bone can also develop. Sarcomatoid carcinoma of thymus is an extremely rare condition with reported incidence of 0.15 case annually in United States and 0.32 case in the Netherlands per 100,000 person years[1,2].

There is a paucity of data on the management of thymic carcinomas, but they are generally treated with multimodality therapy and with similar paradigms to thymomas. Surgical resection, when feasible, is the preferred therapy. There is a greater inclination to use postoperative radiotherapy and chemotherapy because of the higher rate of local and distant relapses; however, the impact of these modalities on clinical outcome is uncertain.

When unresectable, chemoradiotherapy may be an option. The role of radiotherapy for thymic carcinoma is not well defined, again owing to the rarity of the disease and the absence of prospective clinical trials.

Herewith, we are reporting a case of sarcomatoid thymic carcinoma treated at our institute with chemo-radiation.

CLINICAL DETAILS

A 29year old gentleman presented in our outpatient department with history of cough for 4 months, gradually progressive in severity which was associated with mild expectoration and shortness of breath with mild dysphagia. He also gave history of vague chest pain with low grade fever for 4 months. On examination, a right sided cervical adenopathy was detected. Cytology was done from the same and was suggestive of malignancy.

Imaging of the HRCTV thorax showed Mediastinal enhancing soft tissue mass enveloping the Aorta, PA, SVC. Hilar nodes also enlarged on both sides with multiple cyst in liver and kidneys (fig 1).

Core biopsy showed features of Sarcomatoid carcinoma of thymic origin in Mediastinum (fig 2). IHC markers showed Pan CK, Vimentin, CD 34 and SOX 2 positive. SMA, Desmin, ALK 1, CK 5/67, CD 30 negative. Ki 67 was 10%.

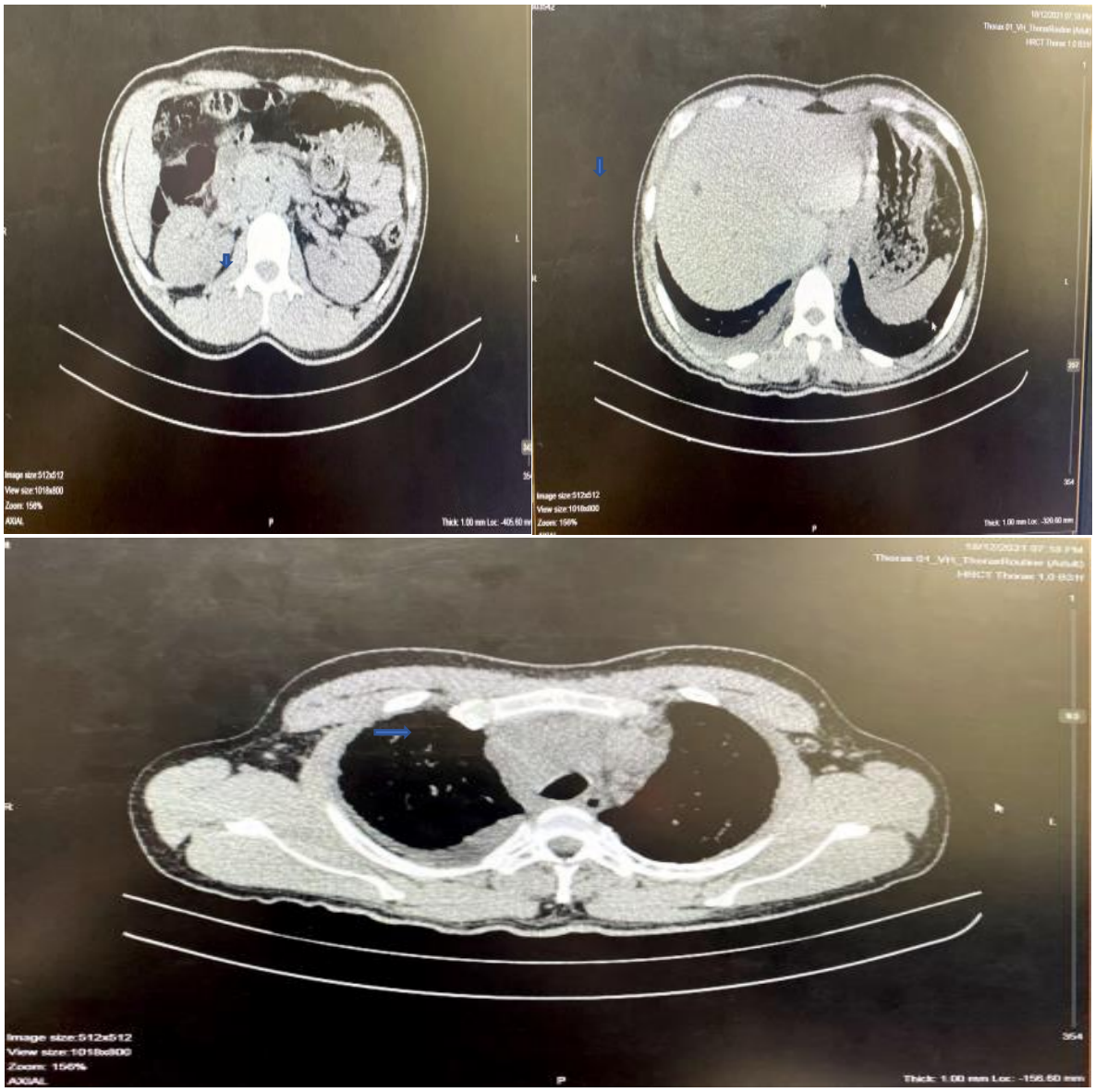

Fig 1. HRCT imaging showing anterior mediastinal mass with multiple cysts in liver and kidney

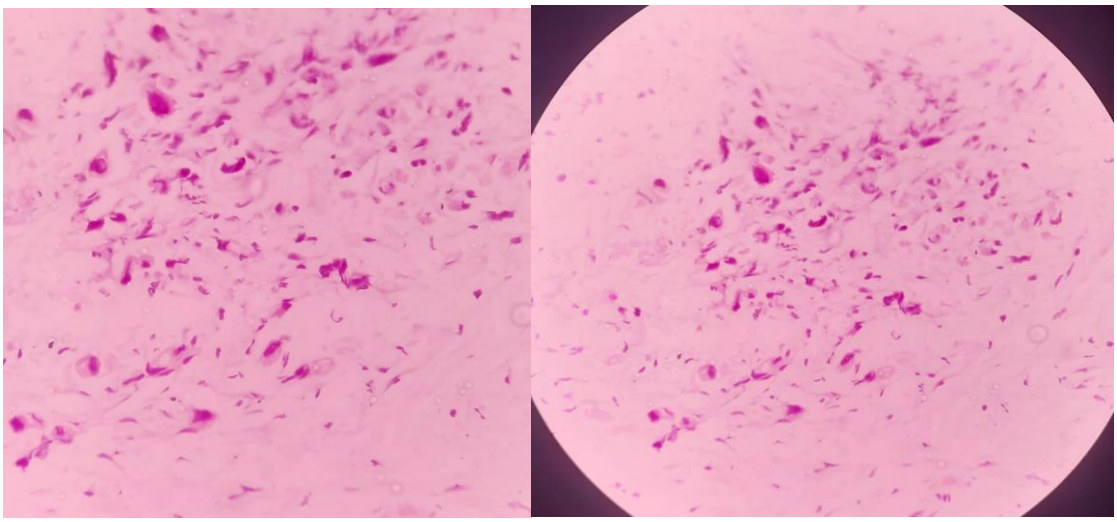

Fig 2. HPE showing Sarcomatoid carcinoma of thymic origin in Mediastinum

The case was discussed in our tumour board and in view of advanced stage it was decided to go ahead with chemoradiation. He was treated with Chemoradiation with tumour dose of 64.8 Gy/36 fractions by IMRT (fig 3 shows the Treatment plan by photon therapy) along with 3 weekly Inj. Etoposide and Cisplatinum Day 1-3 for 2 cycles during

the radiation therapy.At the time of completion of treatment, patient improved symptomatically.

At 3 months post treatment, the patient presented with complaints of headache and blurring of vision. PET CT whole body was done and showed with multiple secondaries in brain, liver, bones. Near complete resolution of the primary lesion in the Anterior Mediastinum was seen (fig 4).

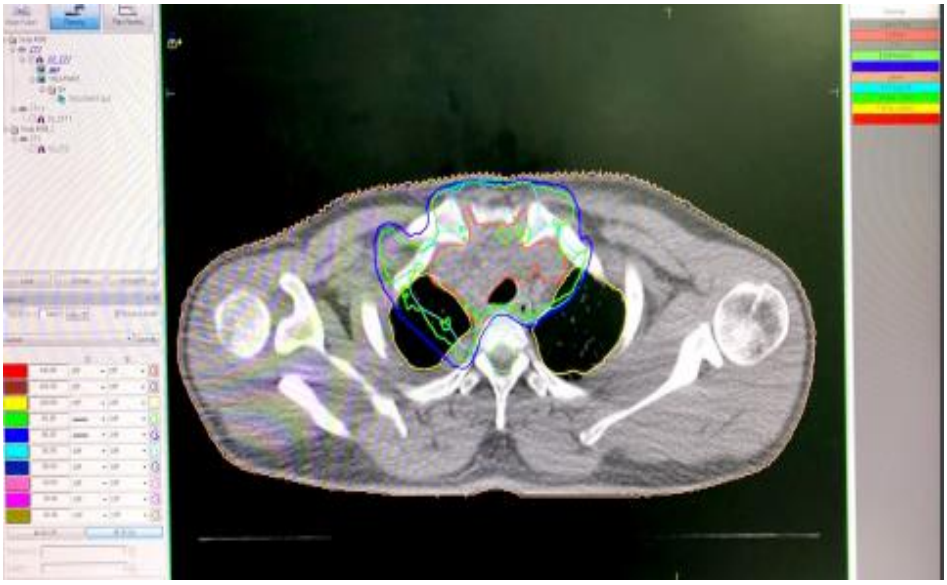

Fig 3. Showing treatment plan by IMRT technique

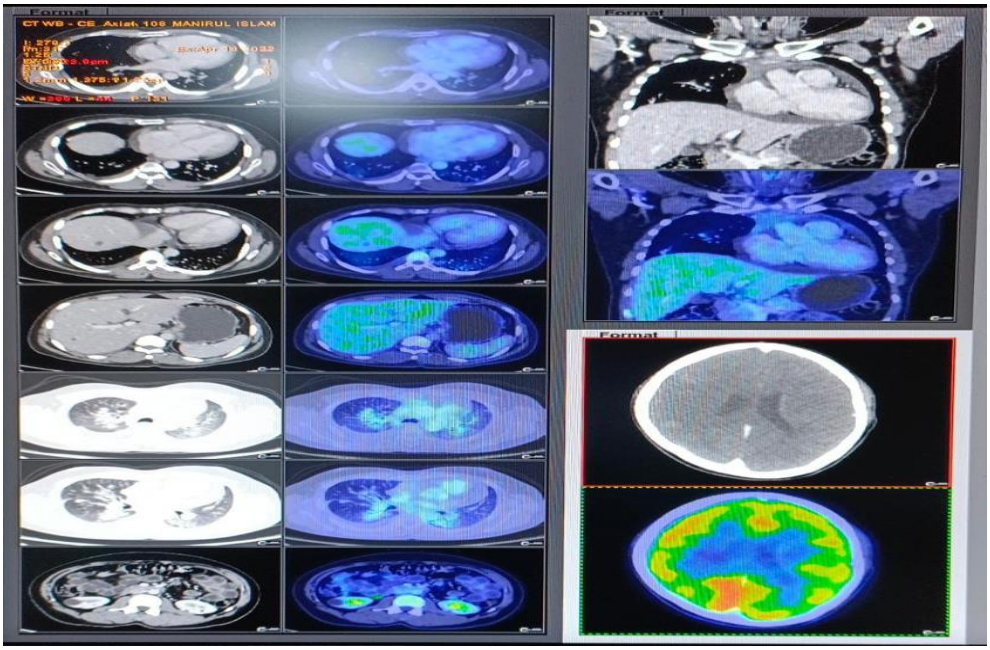

Fig 4. PET CT imaging showing near complete resolution of primary lesion and metastatic lesion in brain on follow up post treatment

Patient was planned for Palliative whole brain RT for 30 Gy /10 fractions. He received the same and was discharged in a stable condition and was advised for further palliative chemotherapy but patient did not come back follow up treatment.

DISCUSSION

Sarcomatoid carcinoma of thymus is an extremely rare condition. Owing to the rarity of the incident case, there is no proper guideline laid down for the management of the tumour. In a case report on Sarcomatoid carcinoma thymus by Mitsuyo Nishimura et al, it was observed that the outcome of the tumour was very poor and the patients died either of local recurrence or distant metastasis at 13 and 28 months. The median survival time of sarcomatoid carcinoma thymus is 24.9 months and 5year survival rate is 42.3% [3].

They usually present as an aggressive tumour presenting with advanced disease at the time of diagnosis. Our patient presented at advanced stage with Mediastinal enhancing soft tissue mass enveloping the Aorta, PA, SVC. It may also arise

from metaplastic thymoma[4]. Diagnosis becomes difficult with a small piece of tissue sample as in a core biopsy sample[5]. The IHC markers usually help to confirm the thymic origin of the tumour. Thymic carcinomas stain positive for Pan CK, p63, CK5/6, PAX8, C-kit, CD5 [6].

There seems to be a trend of better survival with complete or near complete surgical resection of the tumour[6]. Management consists of primary surgery since the role of Radiation and Chemotherapy is not yet established[7]. However there appears to be some role of adjuvant Radiation therapy[8].

The present case presented in an advanced stage involving all the mediastinal structures which have made surgical procedures difficult therefore a nonsurgical approach was taken up. There were no reported cases in literature on definitive chemoradiation. On further follow up 2 months post treatment, PET CT whole body was done and showed near complete resolution of primary lesion.

CONCLUSION

Chemoradiation helps in achieving good response to primary lesion but in view of aggressiveness of tumour biology, aggressive systemic chemotherapy role needs to be defined in the future.

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