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CASE REPORT **OPEN ACCESS**

A MULTIDISCIPLINARY APPROACH OF UNUSUAL SITE LEIMYOSARCOMA WITH 10 YEARS SURVIVAL AND COUNTING

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ABSTRACT

Soft tissue sarcoma (STS)–Leimyosarcoma of the chest wall is a rare entity with dismal survival. Surveillance, Epidemiology and End Result a 5 year survival rate of STS was 81%, 56%, 15% for localised, regional and distant metastasis respectively. Here we present a 31 year man with chest wall leimyosarcoma who is treated with primary surgery and multiple metastasectomy along with six lines of chemotherapy. The outcome of these efforts has resulted in prolong survival with good performance status worth reporting.

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INTRODUCTION

Soft tissue sarcomas (STS) are malignant mesenchymal precursor cell/s that differentiate among one or several lineages, muscle, adipose, fibrous, cartilage, nerve or vascular tissue.(1) They arise in the limbs, followed in order of frequency by the abdominal cavity, thoracic region and head and neck. STS of the chest wall present as painless slow growing masses and account for 0.1–0.15% of all adult malignancy may represent primary or direct invasion of an intrathoracic mass or as metastatic lesions. For most STS of chest wall the primary metastatic site is the lungs via hematogeneous spread while spread to locoregional lymph nodes is rare. (2)

Case report

A 30 year old male with complains of heaviness and swelling in the right side chest associated with occasional pain with no aggravating and relieving factors since June 2011 reported to a local centre.

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A fine needle aspiration showed smears poorly cellular, occasional fragments of fibro fatty tissue possibility of a lipoma with no atypical cells. October 2011 patient underwent excision of soft tissue swelling .The surgical histopathology(HPE) revealed a well encapsulated soft tissue mass 80X50X40mm, microscopically showed proliferation of spindle shaped cells in fascicles with cellular pleomorphism, mitosis and necrosis suggestive of aggressive disease likely high grade leimyosarcoma. The patient was referred to Gujarat Cancer Research institute for further management. Slide and blocks review confirmed the previous pathology finding on which immunohistochemistry (IHC) showed vimentin, actin, desmin positive and S100 negative favouring Leiomyosarcoma. Magnetic Resonance Imaging of right lateral chest wall showed a 31X15X60mm residual lesion at operated site with presence of multiple axillary nodes, largest measures 9X7 mm. In November 2011 he underwent completion local wide excision with axillary lymph nodes dissection. HPE showed no residual tumor and all margins of resection free of any tumor and nodes negative. Patient was kept on regular surveillance thereafter. After 2 year of disease free interval (DFS), patient complained of right hip pain, breathlessness on exertion. In November 2013 A Positron Emission Tomography and Computed Tomography (PET-CT) scan was ordered which showed FDG avid lytic lesion involving superior margin of right acetabulum (SUVmax- 9),

low FDG avid two soft tissue density nodules in anterior segment of right upper and lower lobe of lung.(SUVmax- 2). Biopsy from acetabulum region was done suggesting Metastatic Leiomyosarcoma confirmed on IHC. Patient was offered four cycles of combination of Ifosfamide and Anthracycline which he tolerated well with good radiological response. On March 2014 right lung metastectomy was done followed by Type II right pelvic resection with extracorporeal radiation therapy (ECRT) and plate fixation done in April 2014. Sections from adjacent lung were unremarkable and curettage from ECRT showed no residual tumor. He further received two more cycles of ifosfamide and anthracycline till June 2014 with good tolerance reaching the cumulative dose for anthracycline and was kept on regular follow-up. In March 2015, PET-CT scan showed reappearance of lung nodules for which Pazopanib 800mg was started. In November 2015 PET –CT scan showed absence of FDG avid active disease in the body suggesting complete metabolic response. Pazopanib was continued and he was on 3monthly follow up. In December 2016 PETCT scan showed reappearance of lung lesions which were non FDG avid for which Right lung metastectomy was repeated, HPE: leiomyosarcoma. Post metastectomy imaging was normal. He was continued on Pazopanib as there was no new lesion and previous lung lesion was less than 2cm with good initial response. In August 2017 he developed hypothyroidism, hypertriglyceridemia, mucositis, hypertension and renal dysfunction for which the dose was reduced to 600mg f/b 400mg and stopped in February 2018 and kept on observation.

In April 2018 PET CT showed appearance of new right lobe lung lesion (SUVmax 9.1). He was taken for right lung metastectomy but failed due to a large hard mass medially fixed to the vertebra. In May 2018, 3 cycles of Ifosfamide and Etoposide disease. After 1.5years of progression free survival, he was started on Dacarbazine 6 cycles till May 2020 for appearance of new lung lesions. In June 2020 PETCT suggested complete resolution of pulmonary metastatic lesion. In October 2020 he had PET CT proven relapse in lung for which repeat biopsy for molecular testing was planned but due to inadequate tissue was not possible. He was started on Eribuline but progressed on treatment. In view of long DFI he is rechallenged with Pazopanib, is doing well.

DISCUSSION

Primary soft tissue sarcomas of the chest wall are rare accounting for 6% of all STS and 50% of all malignant tumors arising on the chest wall.[3] The clinical behaviour of chest wall sarcomas is more similar to extremity STS treated similarly. Wide local resection is the standard curative treatment with adjuvant radiation therapy for lesions with close or positive margins. 5-year overall survival (OS) are 63 to 89 percent, outcomes are less favorable with high-grade histology and with incomplete (R1, R2) as compared with complete (R0) resections.[4] Our patient underwent primary R0 resection and had DFI of 2years. Anthracycline- Ifosfamide represents a first-line standard of care with 2- and 3-year OS were 73% and 52% in chemonaive patients. [5] Doxorubicin causes reversible myelosuppression, mucositis, alopecia, nausea and vomiting, cardiotoxicity. 6 cycles was given as first line in metastatic setting with curative intent undergoing metastectomy. Complete pulmonary metastasectomy in a single-institution series of 539 patients undergoing 760 therapeutic-intent

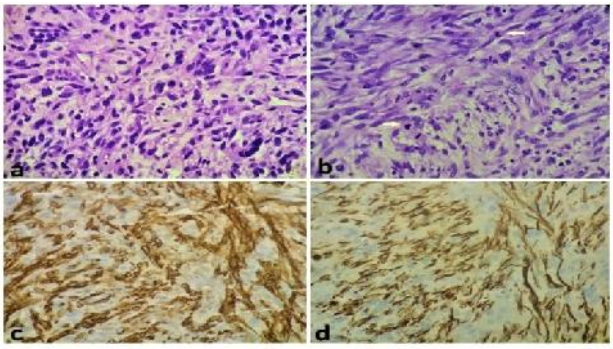

Figure description

a. Tumor cells are arranged in fascicles, having spindle to oval pleomorphic hyperchromatic nuclei and eosinophilic tapered cytoplasm (H&E, 40x) b. Pleomorphic tumor cells with atypical mitotic figures (Yellow arrow) (H&E, 40x) Tumor cells show strong and diffuse expression of SMA (c) as well as Desmin (d) (IHC, 40x) H&E- Hematoxylin & eosin; SMA- Smooth muscle actin; IHC- Immunohistochemistry

pulmonary metastasectomies for metastatic soft tissue sarcoma, the median survival was 33 months and the five-year survival was 34 %.[6] After an R0 pulmonary resection, 74 percent developed a disease recurrence at any site, 63 percent experienced a lung recurrence, and 34 percent had a recurrence that was lung isolated. In multivariate analysis, leiomyosarcoma subtype, primary tumor size ≤10 cm, greater time between primary resection and development of metastases, solitary lung metastases, and minimally invasive resection were associated with a lower risk of death. Our patient had a PFS of 9 months post metastectomy. Pazopanib multitargeted orally active small molecule inhibitor of tyrosine kinases is a subsequent therapy line for who progress on prior chemotherapy. In PALETTE trial, randomized, double-blinded, phase III study median PFS was significantly more in the pazopanib vs. placebo group (4.6 vs. 1.6 months) and benefit was consistent across all histologic subtypes. [7] There was no significant difference in overall survival (12.5 versus 10.7 months, HR 0.86, 95% CI 0.67-1.1).Our patient had a PFS of more than 2 years on pazopanib. The common grade 3 or 4 treatment-related toxicities were fatigue, hypertension, diarrhea, hypothyroidism, azotemia, anorexia, and transient elevations in hepatic functions requiring dose modification also seen. In a series of 86 patients undergoing reoperation for recurrence of pulmonary metastases, the median disease-specific survival was 43 months and the estimated five-year survival was 36 percent.[8] Ifosfamide has antitumor activity against STS, toxicities are severe and dose dependent. Myelosuppression, encephalopathy, hypersensitivity, hemorrhagic cystitis, nephrotoxicity as renal tubular necrosis reported in our case making treatment course stormy. Fourth line Gemcitabine and Docetaxel was administrated to our patient in view of progression, poor tolerance to ifosfamide and had PFS of 1yr 5months. Among the novel agents on further progression our patient could not be put on Trabectedin due to pre-existing renal dysfunction, was given single agent Dacarbazine an alkylating agent as fifth line. He had a progression after 3 months completion of 6 cycles of while on observation and put microtubular inhibitor Eribulin approved for third or greater line therapy in metastatic setting with PFS 2.2months.[9] Our patient developed progression while on treatment with Eribulin and is presently rechallenged with pazopanib in view of long PFS and durable response to treatment, is alive, doing well.

Conclusion

Here we describe multidisciplinary approach in a notorious recurring rare chest wall leiomyosarcoma treated with multiple metastectomy and subsequent lines of chemotherapy culminating in long term survival of more than 10 years, still alive emphasising timely follow-up, diagnosis and treatment offer favourable survival to aggressive high grade malignancy.

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