INTRODUCTION

Primary small cell carcinoma esophagus was first reported by McKeown in 1952. Incidence of small cell neuroendocrine carcinoma oesophagus is rare, and it accounts for 0.5–2.4% of all primary esophageal malignancies. Small cell neuroendocrine carcinoma esophagus is histologically similar to pulmonary small cell neuroendocrine carcinoma. Various methods like surgery, chemotherapy and radiotherapy are used in treatment for small cell carcinoma oesophagus but no optimal treatment has yet been established. We are reporting a case of localized small cell neuroendocrine carcinoma oesophagus treated by chemotherapy and radiotherapy.

CASE REPORT

A 56-year-old female visited to our hospital with a complaint of gradually increasing dysphagia in May 2016. On physical examination, no abnormal finding was detected. Computed tomography (Fig. 1) was done, which was suggestive of asymmetric thickening of a short segment of mid thoracic oesophagus with no mediastinal or hilar lymphadenopathy and no evidence of metastasis in the lungs or liver. Upper gastrointestinal endoscopy showed non-circumferential thickening of mucosa from 26 to 29 cm (Fig. 2). Histopathology report was suggestive of small round cell tumour (Fig. 3). Immunohistochemistry report showed immunoreactive for cytokeratin (CK), Chromogranin A (CGA), Synaptophysin and Ki-67 value 30–40%, suggestive of small cell neuroendocrine carcinoma. All hematological and metastatic work up was normal. She was planned for six cycles of chemotherapy with Inj.Etoposide 100 mg/m² D1-D3 and Inj.Cisplatin 75 mg/m² D1, Q3 weekly. Radiotherapy was added over chemotherapy, starting from 2nd cycle of chemotherapy. Dose of radiotherapy was 45Gy/25#/5 weeks +10Gy/5#/1 week (boost) (5# every week) by 3D CRT technique and six cycles of chemotherapy with Inj.Etoposide 100 mg/m² D1-D3 and Inj.Cisplatin 75 mg/m² D1, Q3 weekly. On follow-up investigations, there was no evidence of local or metastatic disease. She is still disease free after 17 months of diagnosis.

DISCUSSION

Primary small cell neuroendocrine carcinoma oesophagus is rare, highly aggressive and have poor prognosis. Metastasis is present in most cases at
Chemo-radiation in small cell neuroendocrine carcinoma oesophagus: a case report

the time of diagnosis. Small cell carcinoma comprises between 25 and 35% of pulmonary and only about 5% have an extra pulmonary origin. Among the multiple treatment options like surgery, chemotherapy and radiotherapy, there are no established guidelines, due to the rarity of the tumour. Among small cell neuroendocrine carcinoma oesophagus patients, those treated with surgery alone, the result is extremely poor despite of complete resection, due to a high rate of disease recurrence. The median survival was 3.1–20 months. Mitani et al. showed that surgical resection after obtaining a complete reduction of the localized primary esophageal small cell carcinoma by chemotherapy may achieve longer survival. Similarly, in patients with localized disease role of surgery as part of multimodality treatment is controversial, but in some patients long survivals have been reported in those treated with esophagectomy combined with chemotherapy or radiotherapy. A retrospective review was done in 18 patients with small cell carcinoma of the esophagus diagnosed at the Tata Memorial Hospital, Mumbai between 1985 and 2001. 7 of 18 patients (39%) were presented with metastatic disease. 4 patients were treated with surgery, with or without chemotherapy or radiotherapy. 3 patients were treated with chemo-radiotherapy, two patients with chemotherapy alone, and five patients with radiotherapy alone. 4 patients who had a poor general condition were not given any active treatment. The overall median survival of patients was 6 months. Patients treated with surgery and chemotherapy had a better overall survival. Nichols from Memorial Hospital in New York reported small cell carcinoma patients treated with chemotherapy...
alone or with sequential radiation but the overall survival was only 7.5 months. Yamashita et al. studied the effect of chemo-radiation in limited disease small cell carcinoma oesophagus patients treated with four cycles of Etoposide (100 mg/m², days 1–3), combined with Cisplatin (80 mg/m², day 1), plus radiation therapy (50Gy/25#/5 weeks). At the time of analysis, the median follow-up time was 10.8 months (range 4.2–42.8 months) and 21.8 months in five living patients (56%) In a retrospective study of 11 patients of small cell neuroendocrine carcinoma oesophagus, who were treated with radiation therapy and chemotherapy between May 1996 and October 2007, the mean follow-up time was 14.7 months and the median overall survival time of all patients was 13.2 months.

CONCLUSION
Small cell neuroendocrine carcinoma oesophagus is like its pulmonary counterpart. It should be treated like small cell carcinoma lung only. Combined modality therapy using platinum-based combination chemotherapy and radiotherapy appears effective for loco-regional disease. The role of either upfront surgery or after neoadjuvant chemotherapy or radiotherapy or chemo-radiotherapy is not clear, as various studies and case reports are showing variable and non-consistent results.

REFERENCES