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Case Report

Small cell carcinoma of esophagus: Case report

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ABSTRACT

Small cell carcinoma of esophagus is tumor belonging to the family of extra-pulmonary small cell carcinoma. It is a highly aggressive tumor and early diagnosis and treatment is necessary for better prognosis of the tumor. We present a case of 45 year old male diagnosed with primary small cell carcinoma of esophagus and a review of literature of this rare tumor.

1. Introduction

Primary small cell carcinoma of esophagus is a rare tumor. It accounts for less than 2.4% of all esophageal malignancies and very few cases have been reported till date since the first report by Mc Keown.[1] It is associated with a high incidence of metastatic disease at presentation and a poor overall outcome.[2] Clinical symptoms of primary small cell carcinoma of esophagus including X-Ray manifestations and endoscopic observations are very similar to those of esophageal squamous cell carcinoma and adenocarcinoma and thus can only be diagnosed by histopathological examination.[3] Histopathological pathological features are similar to small cell carcinoma of lungs and extra pulmonary small cell carcinomas.[4] Better prognosis is possible with early diagnosis and treatment.

2. Case Report

45 year old male presented with the history of difficulty in swallowing for solid foods for 17 days. The pain is intermittent and aggravated by talking. He also had fever for 4 days. There is history of weight loss 2kg in last 2 weeks. He has h/o gastritis for last 1 year.

Barium swallow was done which showed eccentric filling defect in the thoracic part of esophagus with narrowing of gastro esophageal junction. USG abdomen was normal. CT chest and abdomen showed Ca distal esophagus with involvement of cardia of stomach. The upper GI endoscopy gave an impression of Carcinoma esophagus.

Histopathological examination revealed tissue lined by stratified squamous epithelium which was unremarkable. The sub mucosal area showed tumor cells arranged in solid pattern. The cells show crushing artifacts. The cells were small with dark nuclei and scanty cytoplasm. Immunohistochemistry showed positivity for Chromogranin A and Synaptophysin.

A diagnosis of Small cell carcinoma esophagus was made.

3. Discussion

Squamous cell carcinoma accounts for approximately 90% of esophageal cancer worldwide. In contrast, small cell carcinoma esophagus is an uncommon, rapidly lethal esophageal malignancy with an average survival of less than one year.[1] The incidence ranges from 0.05% to 4.7%.[5]

McKeown first reported two cases of esophageal small cell carcinoma which he observed among more than 9000

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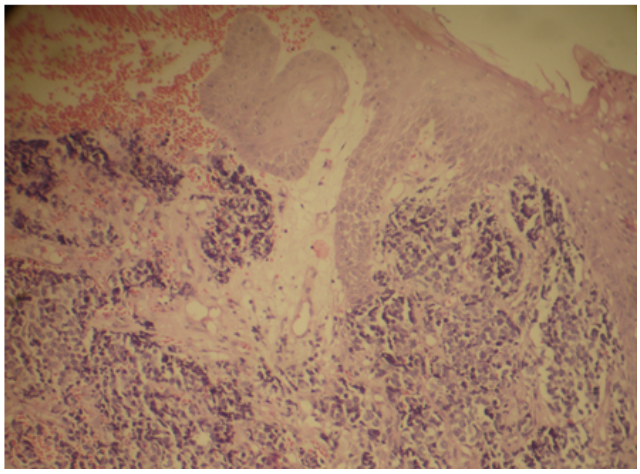

Fig. 1: Normal oesophageal lining with small cell tumor in the sub-epithelium (H&E, 100x)

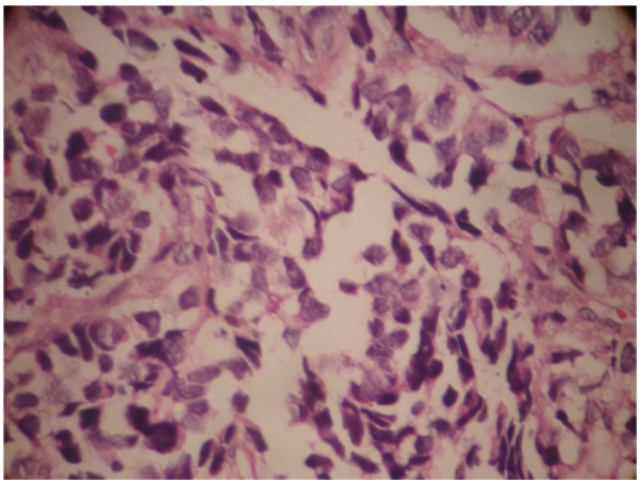

Fig. 2: Small cell carcinoma of oesophagus (H&E, 400x)

autopsy specimens.[6] Since then there were only scattered case reports and a few published series. Fewer than 300 cases have been reported worldwide in literature.

It is a highly aggressive tumor with high incidence of metastasis disease at presentation and is associated with poor outcome.[7] Similar to other esophageal tumors, patients with SCC esophagus present with vague symptoms including constitutional symptoms, weight loss, chest pain or pressure, abdomen pain, dysphagia and odynophagia.[8] Dysphagia is the presenting symptom in most[5](as is similar in our case).

Small cell carcinoma is a rare histological sub type of esophageal cancer.[5] Although most small cell carcinomas are pulmonary in origin, extra pulmonary tumors have been diagnosed in a few cases. Extra pulmonary cases have been reported in larynx, trachea, salivary glands, pancreas, uterine cervix, prostate, skin and oesophagus.[9]

Microscopically, small cell carcinoma is indistinguishable from its counterpart in lung according to histological and immunohistochemical features as well as clinical behavior. According to WHO criteria for small cell neoplasia, the cells small with dark nuclei of round or oval shape and scanty cytoplasm or be larger with more cytoplasm forming solid sheets and nests. Immunohistochemical reaction for neuron specific enolase, synaptophysin, chromogranin and leu 7 are positive and represent useful diagnostic markers. Some cases have been associated calcitonin and ACTH production.[10]

No predisposing risk factors have been identified with small cell carcinoma but some associations with smoking alcohol consumption and Barrett's disease have been made. Mean age of diagnosis is between 40-70years with male predominance. When the disease is suspected, confirmation by histological diagnosis has to be done with adequate staging. Median survival is between 7-19 months.[11]

Primary small cell carcinoma is clinically a very aggressive tumor with and most patients present with an advanced stage and have poor prognosis. Reported cases of small cell cancer esophagus have been treated with various forms of treatment including surgical reaction, radiotherapy, chemotherapy and combinations of therapy but are difficult to compare and assess for efficacy due to small number of patients and the lack of controlled trials. With small cell carcinoma of esophagus, it is almost impossible to get complete healing and local treatment alone is not favorable. It should be treated by multi drug chemotherapy with or without radiation as the first line of treatment. If the general status of patient permits, the regimen of chemotherapy should be continued to achieve a longer disease free survival.[12]

Recent genetic discoveries based on tumor genome sequencing suggest that the Wnt pathway plays important roles in tumor biopsy. Activation of Wnt pathway releases B-catenin which interacts with T cell factor family member to activate the transcription of downstream target genes. R- spondins potently enhance B-catenin signaling and have been implicated in human disease and malignancy. Lgr5 is a receptor for R-spondin and was shown to activate B-catenin signaling when bound to R-spondin and maybe associated with tumorigenesis via the Wnt pathway. Study result suggests Lgr5 protein expression may represent a possible prognostic marker in small cell carcinoma patients.[13]

A better prognosis might be possible with early diagnosis and treatment strategies incorporating chemotherapy along with oncologic radical surgery and/or radiotherapy as part of a multimodality approach. Since treatment protocols are not well established due to the rarity of neoplasm, multi institutional studies are needed to obtain sufficiently large populations for investigations and optimization of therapy of the disease.

4. Source of Funding

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5. Conflict of Interest

The authors declare that they have no conflict of interests.

6. Informed Consent

Informed consent was obtained for the patient for the publication of the case report.

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