Case report

Carcinosarcoma of oesophagus: A case report

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ABSTRACT

Oesophageal carcinosarcoma is a rare type of oesophageal cancer composed of both epithelial and mesenchymal components, occurring with an incidence of about 0.1-1.5 % of all oesophageal tumors. Most of the cases have been reported from Japan. Very few cases have been reported from India. We report a case of 65 year old female who presented with dysphagia and weight loss. Endoscopy revealed a bulky mass and the preliminary diagnosis was oesophageal carcinoma. On histology of the surgical specimen, the tumour revealed both epithelial and sarcomatous malignant cells. A tentative diagnosis of oesophageal carcinosarcoma was made. Immunohistochemical studies showed positivity for both pan-cytokeratin and vimentin thus confirming the diagnosis.

1.Introduction

Carcinosarcoma is a rare malignant neoplasm characterised by the presence of both epithelial and mesenchymal components. The incidence is about 0.1-1.5 % of all oesophageal tumours [1]. This mixed type of tumour has been reported in various organs, including uterus, vagina, lungs, oral cavity, larynx, thyroid, urinary tract, and oesophagus. The clinical features and risk factors are similar to that of oesophageal squamous cell carcinoma, but prognosis is better [2].

2.Case Presentation

A 65 year old female presented with complaints of progressively worsening dysphagia and weight loss for 3 months. Upper G.I. endoscopy revealed an obstructing mass in the oesophagus. Biopsy showed only malignant epithelial cells and the preliminary diagnosis was oesophageal squamous cell carcinoma. Further CT scan studies showed no radiological evidence of metastatic disease. The surgical specimen was sent for histopathological examination.

The received specimen of oesophagus was 14cm long. 4.5 cm from the distal end was a polypoidal lobulated ovoid soft tissue mass adherent to the oesophagus with a short pedicle. Cut section showed a fleshy appearance. The specimen was adequately fixed in 10% formalin. Representative sections were taken, processed and embedded in paraffin wax. The sections were stained with hematoxylin and eosin.

Multiple histopathological sections of the mass studied, revealed a tumour composed of two different elements – sarcomatous and epithelial. The sarcomatous component showed sheets of plump malignant spindle shaped cells with vesicular hyperchromatic nuclei showing prominent nucleoli [Figure 1]. The epithelial component was made of undifferentiated epithelial cells with atypical nuclei [Figure-2]. Mitosis was high (2-4/high power field). Large area of necrosis with infarction was also seen. Rest of the oesophagus appeared normal. Resected ends appeared normal. Histolopathological diagnosis was carcinosarcoma of the oesophagus.

On immunohistochemistry, the sarcomatous cells stained positive for vimentin [Figure 3] and the epithelial cells for pan-cytokeratin [Figure 4], thus confirming the diagnosis of Carcinosarcoma.

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FIGURE 2: Epithelial areas composed of undifferentiated cells and mitotic figures admixed with sarcomatous spindle cells (H&E x 100)
Inset – higher magnification (x400)

FIGURE 3: Sarcomatous areas showing vimentin positivity (x400)

FIGURE 4: Epithelial areas showing pan-cytokeratin positivity (x400)
3. Discussion

Carcinosarcoma is a rare malignant neoplasm characterised by the presence of both epithelial and mesenchymal components. It has also been referred to by other names, including spindle cell carcinoma, pseudosarcomatous squamous cell carcinoma, polyoid carcinoma, and squamous cell carcinoma with a spindle cell component [3]. This mixed type of tumour has been reported in various organs, including uterus, vagina, lungs, oral cavity, larynx, thyroid, urinary tract, and oesophagus. Most of the cases have been reported from Japan. Two theories evolved to explain the development of oesophageal malignancies. Most of the cases have been reported from Japan. Two theories evolved to explain the development of oesophageal malignancies. The theory suggests that 2 distinct malignancies coexist and then collide and intermingle. The second theory suggests there is a reciprocal, neoplastic induction of the epithelial and stromal elements [4].

Like other oesophageal cancers, oesophageal carcinosarcoma is more prevalent in men [5], especially those with a history of tobacco and alcohol use [6]. It is characterized by rapid growth and complaints of dysphagia and weight loss. On endoscopic examination, the lesion appears bulky, polyloid, gray-white mass with smooth, lobulated or scalloped margins. The mass is most commonly located in the middle third of the oesophagus, followed by the lower third and the upper third [7]. It is difficult to distinguish this lesion from other types of oesophageal tumours radiographically. Endoscopic biopsy often shows the superficial epithelial cell component but not the deeper sarcomatous cell component. The final diagnosis depends entirely on the surgical pathology of the mass.

On histopathology, the epithelial component can be squamous cell carcinoma or adenocarcinoma. It stains positively for epithelial markers such as cytokeratin and EMA [8]. The pleomorphic sarcomatous cell component is composed of spindle cells growing in fascicles in a whorled pattern, sometimes exhibiting differentiation toward cartilage, bone, or skeletal muscle and stains positively for collagen, smooth muscle actin and vimentin [5,6].

The prognosis of sarcomatoid carcinoma is much better than that of common squamous cell carcinoma because they tend to grow into the lumen rather than into the wall and they typically invade no deeper than the lamina propria or the submucosa [2,4]. McCort [9] concluded that the carcinosarcoma component of the tumor usually being at an early stage at the time of diagnosis, have a lower incidence of lymph node metastasis. In addition, sarcomatoid carcinomas do not invade early in their course and have a lower tendency to metastasize. Finally, because of their exophytic growth, sarcomatoid carcinomas become symptomatic early in the course of the disease compared with typical squamous cell carcinoma. Both the epithelial component and the sarcomatous component can spread through direct invasion or distant metastasis through lymphatic or hematogenous route to liver, brain, lung, and bone. Recurrence due to hematogenous metastasis is more frequent in carcinomasarcoma than in oesophageal squamous cell carcinoma [10]. Identifying the differences in biological characteristics of the tumour might help guide additional therapy of recurrent cases and improve the prognosis of patients [8].

A rare type of oesophageal cancer is oesophageal carcinosarcoma, and it is unique for being not purely of a single cell type, but instead mixed with both epithelial and mesenchymal components. Its characteristic rapid growth into an exophytic mass leads to symptoms of dysphagia and weight loss. Treatment is primarily surgical resection. Diagnosis is based entirely on the histopathology of the resected specimen, which will reveal the dual population of cells. Immunohistochemistry is essential for the confirmation of diagnosis. Prognosis is comparatively better than the conventional oesophageal squamous cell carcinoma and early detection helps in improving the survival rate.

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References

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