Primary malignant carcinoid tumor of the esophagus

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Primary malignant carcinoid tumor of the esophagus is exceptionally rare. We report a case of primary malignant carcinoid tumor of the esophagus in a 62-year-old man, which was suspected to be leiomyoma or leiomyosarcoma on barium esophagogram. The final diagnosis of malignant carcinoid tumor was made on histopathological and immunohistochemical studies of surgically operated specimen. The patient remains in clinical remission 80 months following surgery and post-operative radiotherapy. [Turk J Cancer 2000;30(1):40-43]

Key words: Esophagus, malignant carcinoid tumor

Carcinoid tumors arise from the enterochromaffin cells (EC), which are scattered throughout the body but occur primarily in the submucosa of the intestine and main bronchi. They have been classified as APUDomas. Approximately 85% of the carcinoid tumors develop in the gastrointestinal tract, 10% are found in the lungs and the rest in other organs such as larynx, kidney, ovary, prostate, skin etc. The most frequent location in the gastrointestinal tract is appendix followed by rectum and ileum (1).

Case Report

A 62-year-old man presented with a 3-month history of progressive dysphagia with solid food and a weight loss of seven kg. There was no relevant past medical history. His general physical examination was normal. Clinically there was no lesion in his oropharynx or larynx. There was no cervical or supraclavicular lymphadenopathy.

Barium esophagogram demonstrated widening of the contrast column with a large eccentric filling defect with ill-defined margins on the posterior wall, mainly on left side in the lower 2/3 of the esophagus (Figure 1). The cardioesophageal junction was normal. The radiological features were suggestive of an intramural mass with possibility of leiomyoma or leiomyosarcoma. Chest X-ray and abdominal ultrasonography were normal. Endoscopic examination of the esophagus revealed a friable mass beginning at 28 cm from the incisors and
occupying 3/4 of the lumen. Endoscopic biopsy was non-contributory as most of the tissue submitted for histology was necrotic with acute inflammatory exudate. The patient was subjected to surgery. Frozen section from the esophageal growth was reported as malignant tumor. Three staged esophagectomy with partial gastrectomy and esophagogastric anastomosis was performed.

Pathological findings: The resected specimen contained a fleshy growth 75 x 55 x 25 mm in size at middle and lower part of the esophagus, mainly on posterior wall. Tumor was mainly submucosal. The outer surface of the tumor was grey-brown and shiny. At some places tumor was capsulated. Most of the tumor was irregular with few areas of hemorrhage and necrosis. On cutting, the cut surface was grey white, smooth and firm. Microscopic examination revealed malignant cells mainly showing trabecular, solid and acinar pattern with areas of hemorrhage and necrosis. The cells showed peripheral palisading. The cells were anaplastic, moderate to large in size having round to oval nuclei. Atypical mitosis, moderate pleomorphism and fair amount of eosinophilic cytoplasm was also noted (Figures 2 A,B). The cardioesophageal junction and the resected part of the stomach were uninvolved. Both proximal and distal margins of
resection were free of tumor. Seven paraesophageal nodes embedded in the fibro-fatty tissue and one of the celiac nodes removed at surgery showed evidence of metastatic tumor (figure 2 C). Special stains: Mucin stains e.g. PAS, PAS with D and alcian blue were negative. With grimelius stain almost all tumor cells showed intracytoplasmic black granules consistent with argyrophilia. Immunohistochemical tests: many tumor cells showed positive reaction for CK and CK-19 antigen. Chromogranin was sparse but definitely positive. Based on these findings a final diagnosis of malignant carcinoid tumor of esophagus with nodal metastases to paraesophageal and celiac nodes was made. His post-operative urinary 5 hydroxy indole acetic acid (5-HIAA) was normal. The patient was subjected to a course of post-operative radiotherapy (dose 50 Gy/25 fraction/5 weeks) encompassing the primary site and the drainage area by two parallel opposed anterior and posterior fields. The patient currently remains alive and disease free 80 months following his treatment.

Fig. 2 (A): Photomicrograph of section of the esophagus showing submucosal carcinoid tumor (H&E stain, scanner view); (B): Photomicrograph of esophageal carcinoid showing solid pattern of growth and pleomorphic large to oval tumor cells with eosinophilic cytoplasm (H&E stain X40); (C): Low power view of the celiac node showing metastatic carcinoid tumor (H&E stain X10)
Discussion

Carcinoid tumors of the esophagus are extremely rare. So far, only five cases have been reported in the English literature (2-6). The present patient is only sixth case in the literature and the first being reported from India. Due to the rarity of these lesions, they practically never enter into the differential diagnosis of esophageal malignancies. In general, carcinoid tumors are seen at any age ranging from 10 to 90 years, though majority are seen in sixth and seventh decades. All the reported cases of esophageal carcinoid were elderly male patients. Their lesions were located in mid and lower esophagus similar to the present case. Our patient was a 62-year-old male. Clinical symptoms, radiological features and endoscopic findings were not conclusive in the patient presented in this report. His barium esophagogram revealed an intramural lesion similar to rather more commonly known, though rare lesions in this location e.g. leiomyoma and leiomyosarcoma. The correct diagnosis could be made only after histopathological and immunohistochemical examination of the tissue specimen.

Though the mainstay of the treatment of carcinoids is surgery, the present patient was also subjected to post-operative radiotherapy due to nodal metastases. He remains without evidence of disease more than six and half years post-treatment.

In summary, though clinical features and radiological findings of carcinoid tumor of esophagus were not specific in the present case, the possibility of an intramural filling defect being due to an esophageal carcinoid tumor should be considered in differential diagnosis of esophageal malignancy.

References