

Verrucous Squamous Cell Cancer of the Esophagus: A Case Report

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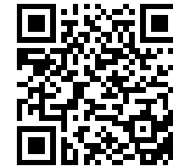
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Abstract

Verrucous Cancer of the esophagus is a type of squamous cell carcinoma and is associated with a chronic inflammatory process in the lower esophagus usually secondary to gastro-esophageal reflux disease (GERD). Mostly it is a local disease process but sometimes it can invade distant structures also. We report a case is a 62-year-old female patient who presented with marked weight loss, anorexia, and difficulty to take solid food. On Upper GI endoscopy there was a warty appearing irregular mass at the mid to distal esophagus. Superficial multiple biopsies were taken but turned negative for malignancy and re-endoscopy after a few days with deep multiple biopsies revealed the diagnosis of verrucous carcinoma on histopathology. The patient was having multiple co-morbid conditions like hypertension, diabetes mellitus, and early nephropathy along with infiltration of malignancy in surrounding structures on CT scan chest. Therefore a decision was taken to place an esophageal stent by upper GI endoscopy followed by chemotherapy and radiotherapy by the oncology department. Patient symptoms were much relieved after 4 months of chemotherapy and radiotherapy and the patient is still on regular follow-up in medical OPD. Chronic inflammation due to gastritis and esophagitis is the main risk factor for verrucous carcinoma of the esophagus. Although surgery is the treatment of the choice for the local and early disease but as in our case disease was already spread palliative stenting followed by chemo-radiation is the best possible option that can be offered.

Keywords: Verrucous carcinoma, Esophageal stenting, Esophageal carcinoma, Endoscopic Ultrasound.

Introduction

Verrucous cancer of the lower esophagus is a very rare form of malignancy mostly of squamous cell origin. A varicoid carcinoma is a slowly growing, well-differentiated but very rare type of squamous cell carcinoma. On Upper GI endoscopy, this looks like a mass that has an irregular, exophytic, and warty appearance.¹ Due to its morphological nature it is very difficult to diagnose it on superficial biopsy samples due to nonspecific histological findings. So, its diagnosis needs a high index of suspicion and most of the time a deep biopsy sample is required with upper GI endoscopy or sometimes with endoscopic ultrasound. Risk factors for this type of malignancy are chronic smoking, gastroesophageal reflux disease (GERD), chronic alcohol use, human papillomavirus (HPV), achalasia cardia, and some other chronic inflammatory conditions.² Superficial mucosal biopsies of cancer mostly shows chronic inflammation often missing the high-grade dysplasia leading difficulty in diagnosis. Incidence of verrucous cancer is higher in males as compared to females between the ages of 35 to 80 years. To date, only 45 to 50 cases of this malignancy are reported in the literature.¹ We reported this case of varicoid carcinoma of the esophagus with mild GERD symptoms as a risk factor and diagnosis was done on deep mucosal biopsies of the lesion.

Case Report

A 62-year-old lady belonging to a rural area, presented with significant weight loss of about 11 kg along with dysphagia, epigastric pain, and retrosternal chest discomfort especially after taking meals for the past 6 months. She was having chronic hypertension for the last 10 years taking regular treatment and well-controlled type 2 diabetes mellitus for the last 14 years taking regular treatment with moderate control and early nephropathy with mild elevation in renal function tests. She gave a history of off-and-on huqqa smoking with mild symptoms of gastro-esophageal reflux for the last 2 years. Her general physical examination showed no gross abnormality. Her laboratory investigations showed mild elevation in urea and creatinine levels, normal LFTs, and blood Hb of 10.5 g/dL (MCV 74.4 fl). The rest of the laboratory examination was normal. Her ultrasound examination along with a CT scan of the abdomen and pelvis turned out negative for any gastrointestinal or pelvic

malignancy. She was advised barium swallow and meal which showed an irregular mass involving the mid-esophagus with an appearance of stricture in the proximal part of the esophagus (Figure 1). On upper GI endoscopy, there was a warty appearance of mass and on endoscopic ultrasound, it was a solid tumor about 4.5 cm in dimension maximum at the mid esophagus and infiltrating the surrounding structures. (Figure 2)

Multiple samples of the endoscopic biopsy were taken and turned out to be non-specific inflammation with normal squamous epithelial cells, parakeratosis, ulceration, and some atypical cells. A repeat endoscopic biopsy was taken with samples from deep mucosa with jumbo forceps which on histopathology confirmed the diagnosis of verrucous carcinoma of the esophagus. The bone scan was negative for any evidence of metastasis. A Computed Tomography scan of the chest revealed mediastinal lymphadenopathy nodes and infiltration in surrounding structures. Opinion from a surgical specialist was taken but surgery was contraindicated due to local metastasis. The patient had an esophageal stent placed by upper GI endoscopy (Cook Medical fully covered 23 mm wide and 100 mm long). The patient was referred to an oncologist who planned for a schedule of chemotherapy followed by radiotherapy and is still on follow-up in the medical and oncology outpatient department for chemotherapy and radiation.

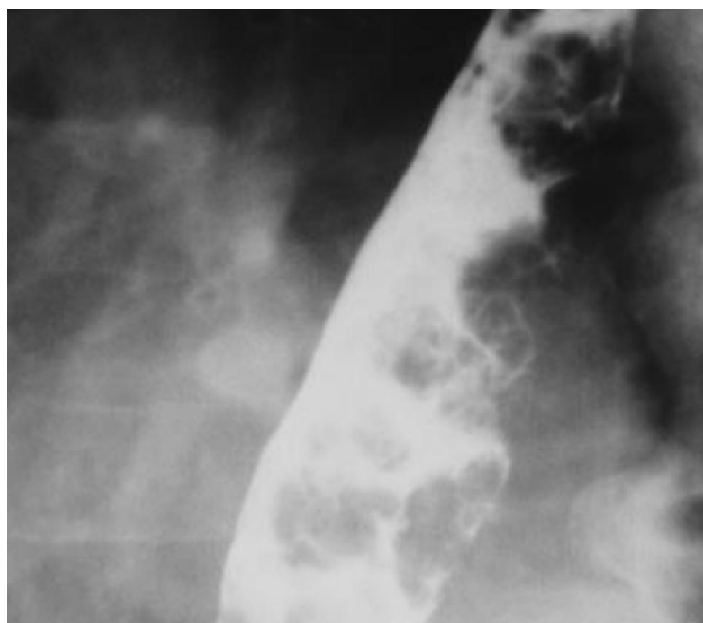


Figure 1: Barium swallow of esophagus showing an irregular mass in the mid esophagus.

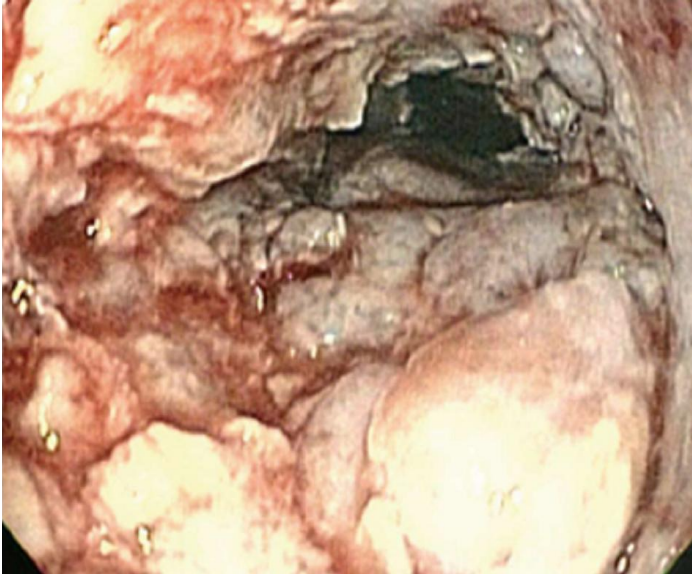


Figure 2: Upper GI Endoscopy showing an intraluminal mass in the esophagus with an irregular warty appearance

Discussion

Varicoid carcinoma is a rare slow-growing variant of squamous cell carcinoma and is seen in many areas like the oro-pharyngeal pouch, over glans penis, scrotal area, female genitalia, endometrium, urinary bladder, anorectal region, and at the sole of foot but among all these sites it is rarely seen in esophagus.³ First ever case of such cancer in the esophagus was reported by Minelly in 1967 and since then only about 50 cases have been reported in the literature.⁴ So far no case has been reported in local areas of India and Pakistan. Risk factors for this kind of malignancy are chronic inflammation which may be due to any precipitating cause like smoking, alcohol ingestion, hiatus hernia, achalasia cardia, chronic esophagitis due to any cause or some corrosive intake.⁵ Some cases have reported association with the Human Papillomavirus.⁶ It has been well established that anti-acid therapies decrease tumor size and prevent it from making growth to a polypoid appearance. So far seen in literature incidence of this cancer is high in the male population (male to female ratio 2:1 approximately) and at ages ranging from 36 to 79 years (mean age is 61 years).⁷ The most alarming presentation of varicoid cancer is symptoms of dysphagia to solids and significant weight loss as was also found in our case. Upper GI Endoscopy may show variable findings like exophytic, wart-like, velvety, papillary, and spiked or cauliflower-like appearances.³ This tumor is mostly

located in the lower esophagus in 70% of cases, the middle part in 23% of cases, and in the upper esophagus in about 7% of cases.⁸ A definitive diagnosis is mostly done by either endoscopic guided deep biopsy of mucosa or Endoscopic Ultrasound-guided biopsy or sometimes post-surgical evaluation of samples taken during surgery.⁹ This cancer morphologically may vary from acanthosis, hyperkeratosis, parakeratosis, leukoplakia, and papillary hyperplasia to varicoid carcinoma.¹⁰ Superficial biopsy of lesions may be negative or show non-specific acanthosis, hyperkeratosis with some chronic inflammation so making it difficult to diagnose these cancers.¹¹ Therefore case of negative biopsy needs a high index of suspicion and a repeat biopsy should be done as it was in our case. In a study, an endoscopic biopsy was positive in 46% of cases while the rest of the carcinomas were diagnosed either after surgery or by the use of EUS.¹² Endoscopic Ultrasound is a highly sensitive imaging modality and very useful in diagnosing these esophageal cancers. EUS can also measure the extent of invasion along with associated lymphadenopathy needed for staging of cancer. Endoscopic ultrasound-guided tunnel biopsies of tumors are very useful as seen in some case reports in the literature.⁷

Even though this kind of tumor is slowly growing and there is a higher degree of differentiation, it has got a very bad prognosis and most of the time the diagnosis is much delayed than the onset of the symptoms and signs. Mostly the morbidity and mortality are mainly related to early local invasion or some surgical complication. Although this tumor can spread locally to the lungs, bronchi, pleura and can form fistulas but distant metastasis is rarely reported in the literature.⁵ If the tumor is diagnosed at an earlier stage, it can be surgically removed as polypectomy or mucosal resection from the esophagus but as the disease is advanced esophageal stent placement or an earlier PEG tube placement if possible are alternative options. As these tumors are rare so far, no clear data is available to see the effectiveness of chemotherapy or radiation therapy. There is a need for long-term follow-up in these patients to see the exact outcome.

Conclusion

A varicoid carcinoma of the esophagus is slowly growing, well-differentiated but a rare form of squamous carcinoma variant associated with local disease spread. On upper gastrointestinal endoscopy, it looks like a projectile warty irregular mass mostly

very difficult to diagnose by superficial biopsy because of negative or non-specific histology. Therefore, it requires a high index of suspicion with deep mucosal biopsy with upper GI endoscopy or endoscopic ultrasound. Early cases can be treated surgically but the advanced disease can be considered for esophageal stent placement or other conservative measures.

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