Black Esophagus: A rarely diagnosed case of Acute Esophageal Necrosis at Autopsy.

Abstract:
Acute esophageal necrosis (AEN) also referred as ‘black esophagus’ or ‘acute necrotizing esophagitis.’ It is a rare clinical disorder, but the mortality arising out of it is still a rarest entity. Some cases were diagnosed in hospital setup by endoscopic examination in admitted patients, but very few cases were autopsied and described in medical literature till date. We have carried out an autopsy of a deceased found on footpath. The autopsy examination reveals the findings of death due to acute esophageal necrosis and those findings was confirmed by histopathological examination. The present case is taken for discussion to enlighten the rare disease ‘acute esophageal necrosis’ and to discuss the profile of the disease.

Key words: Acute esophageal necrosis, Black esophagus, Acute necrotizing esophagitis, Acute inflammatory cells.

Introduction:
Acute esophageal necrosis (AEN) is commonly referred as ‘black esophagus’ or ‘acute necrotizing esophagitis’ is very rarely diagnosed condition. It was firstly described in postmortem case by Brennan in 1967, this was the only case described in pathology literature in postmortem examination till 2011 when another such case was diagnosed by Altenburger DL while doing Postmortem examination. Goldenberg et al found the first case in 1990 while doing endoscopic examination in a hospitalised case.
It is stated that less than hundred cases of esophageal necrosis syndrome was described in literature.\textsuperscript{4} Death due to esophageal necrosis syndrome occurs in less than 6\% cases, but if the comorbidities exist it will raise the morbidity as high as 32\%.\textsuperscript{5} Gross macroscopic and histopathological examination combinely gives good results in determining and confirming the diagnosis of disease condition.

Most widely postulated mechanism for this disease is mucosal ischemia\textsuperscript{3,6,7} seen in hemodynamic compromise conditions, hypotension, and corrosive injury from reflux of gastric contents and gastric outlet obstruction and decreased function of mucosal barrier systems and reparative mechanisms present in malnourished and debilitated physical states, consumption of large amount of alcohol in short time, vasculopathy, gastric volvulus and malignancy.\textsuperscript{8} The disease is commonly seen in male population of older age with male: female ratio 4:1, and mean age for the disease is 68.44 years.\textsuperscript{6}

**Case report:**

The corpse of unknown male victim aged about 50 years was brought to mortuary of forensic medicine department for medicolegal postmortem examination. The deceased was found on a footpath at early morning; the police officer held spot inquiry and brought to forensic medicine department for autopsy to ascertain the cause of death, any evidence of poisoning or any unnatural means that contributed in the process of death. Along with this, the postmortem in unknown bodies carried out to preserve the samples such as blood, piece of bone or tooth for DNA analysis, which help to find the relatives or kin of the deceased.

**Examination:**

**External examination:**

On examination it was a male corpse of aged about 50 years, weight 40 kg and stature about 165 cm. wrapped in bedsheet and dirty mud stained cloths. Built was thin and poorly nourished, skin is thin paper like, dry, inelastic, pale, and cold, Eventually, the fatty tissues in the facial region is lost, the cheeks look hollow and the eyes sunken. Hairs were dry and sparse, plucked out easily. No external injuries were seen over the body.

**Internal examination:**

Internal examination shows the Blackish discoloration in mucosal area of distal part of esophagus, with sharp demarcation of gastrointestinal border. The mucosal area shows the ischemic necrosis and sloughing (Fig.1). The gastric mucosal surface and rugae were normal but areas of gastritis were seen at places. (Fig 2). Stomach contains about 50 cc brownish fluid with sour alcoholic smell. Evidence of cerebral and pulmonary edema was present. The blackish part of stomach i.e. distal part of esophagus, and all the organ pieces preserved for histopathology.
Histopathological findings:

The histopathological examination of distal esophagus shows, necrosis of mucosal surface, sloughing of mucosa at places, congestion of mucosal surface at places, edema of mucosal and submucosal layers, and infiltration of acute inflammatory cells was seen in submucosa, muscularis propria and muscular layer of esophagus (fig.3). On histopathological examination, brain shows cerebral edema with petechial hemorrhages, pulmonary edema. Liver shows fatty infiltration, and kidneys within normal limit.
Discussion:

Esophagus is long muscular tube that connects the pharynx to stomach and extends from lower border of the cricoid cartilage to cardiac orifice of the stomach. Histologically, the esophagus has the four concentric layers of mucosal layer, submucosal layer, muscular layer and adventitial layer. Three portions of esophagus derive its blood supply from various branches of arteries. The cervical portion is supplied by the inferior thyroid artery, the thoracic portion is supplied by bronchial and esophageal branches of the thoracic aorta and the abdominal portion is supplied by ascending branches of the left phrenic and left gastric arteries. When the factors that causing injury to this highly vascular structure leads to ischemic changes that causes death of cells at cellular and molecular level, and also reactive oxygen metabolites formed as a result of reperfusion injury and leukocyte migration may directly and indirectly damage important mechanisms of cellular viability and function, which results in cellular lysis and death.

In present case the body was found on footpath and most probably he was a beggar by appearance. When we consider the possible etiology of disease in present case, it might be due to malnutrition and debilitated status of victim, reflux of acid from stomach due to hyperacidity, which is supported by the findings of gastritis at mucosal surface of stomach, and sour smell like alcohol from stomach. All these factors fit into the etiology of the acute esophageal necrosis.

Two autopsy series from the United States reported zero cases in a series of 1000 adult autopsies and in France 0.2% cases in 3000 autopsies. And two retrospective series that reviewed the findings in more than 100000 endoscopies estimated the incidence at approximately 0.01% (12 patients), and at another instances retrospective analysis of 10295 endoscopies has shown the incidence of 0.28% (29 patients). Zacharia GS et al published a case of black esophagus in 2014 of 62 years old man while doing endoscopy in hospitalized case. Altenburger DL et al published such a case in 2011 of 45 years old man during postmortem examination and also suggested that Positive periodic acid–Schiff and negative iron stains suggest that the pigment is lipofuscin, likely secondary to ischemia.
Conclusion:

Acute esophageal necrosis or black esophagus is a rare disease, and mortality arising out of it is still rarest entity. The various etiological factors contribute for the disease, but in this particular case malnutrition, debilitated condition of patient, alcoholism was the triggering factors. One should take assistance of histopathological examination along with macroscopic findings to establish the proper diagnosis of acute esophageal necrosis.

Compliance with Ethical Standards

Conflict of Interest: The authors declare that they have no conflict of interest.

The study was not funded by any source.

This article does not contain any studies with human participants or animals performed by any of the authors.”

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