

## Case Report:

# Angiokeratoma-like Idiopathic Oesophageal Ulcer, A rare case of dysphagia in HIV female patient, A Case Report

Izzaddinn Elawad <sup>1</sup>, Moawia Elbalal Mohammed <sup>2</sup>, Montasir Elamin <sup>3</sup>, Ahmed Abdallah Mohamedani <sup>4</sup>, Hyder Mergani <sup>5</sup>.

1-Assistant professor of internal medicine, department of medicine, faculty of medicine, University of Gezira.

2- Professor of internal medicine, Gastroenterologist, department of medicine, faculty of medicine, University of Gezira.

3. MD Internal medicine S.M.S.B, Sudan.

4. Professor of pathology , department of pathology , faculty of medicine , University of Gezira .

5- Assistant professor of internal medicine, department of medicine, faculty of medicine, University Tabuk.

Corresponding Author : Dr . Izzaddinn Elawad , department of medicine , faculty of medicine , University of Gezira .Wadmedani, Sudan

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### Abstract:

A female patient with ADIS related complex and symptoms of dysphagia was examined endoscopically. Two kissing giant oesophageal ulcers were found in her mid-oesophagus. Histopathology revealed that they were angiokeratoma –like ulcers. This case is being reported for its rarity.

**Keywords:** ADIS- related complex, dysphagia, giant esophageal ulcers.

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### BACKGROUND:

The esophagus of the immunocompromised host is the potential target for a variety of pathogens <sup>[1]</sup>. Symptomatic oesophagitis with ulceration in patients with acquired immune deficiency syndrome (ADIS) has been well described in monilia infections, as well as cytomegalovirus (CMV) <sup>[2,3]</sup>. Idiopathic ulcers became almost as frequent as CMV ulcers <sup>[4]</sup>.

### CASE REPORT

A 34 years old female reported to us with a three-month history of dysphagia for solids. She had numerous complaints of epigastric pain as well as right anterior sharp chest pain. She said loose motions that were on – off over the same period, and she has a considerable weight loss.

Her social history revealed that she is a widow and a mother of 6 kids; the youngest one died three years ago with HIV disease and active pulmonary tuberculosis. Her husband died four months later with the same disease at Saudia Arabia. During her clinical work-up, she started to have more frequent motions, increase the difficulty in swallowing and nocturnal fever.

Clinical examination revealed a young tall wasted and slightly pale female. There were three purple papules on her left cheek. Examination of her hard palate showed another purple papule. Her B.P was 110/70 mmHg, and her pulse was 100/minute. Her cardiovascular, chest and central nervous system were all normal. There were no visceromegaly or lymphadenopathy.

Laboratory studies revealed a normal hematocrit, hemoglobin, white blood cell count, differential, platelet, and red blood cell morphology. ESR was 90 mm/hr, LFT, RFT, and lipid profile were all within normal limits. Stool examination showed bacteria. Her chest x-ray was normal, and her sputum for Zn-stain was negative for tubercle bacilli.

The alpha-galactosidase A- level in blood was found to be 16.83 mU/mg with a normal range of (9.60-42.2). HIV (ELISA) was reactive. Oesophagogastroduodenoscopy was done that revealed two giant kissing ulcers at her mid-oesophagus, with multiple purple papules at the pyloric antrum (see fig.1& 2).

The histopathological examination of the oesophageal biopsy showed dilated blood channels, running intramucosal and in the submucosa, that resemble angiokeratoma figure (3). The gastric biopsy showed dilated vascular channels below the surface. No typical features of Kaposi's sarcoma were seen in either biopsy. Biopsies taken from lesions on her left cheek showed characteristic features of Kaposi's sarcoma.

From the above mentioned scenarios the patient was classified as stage IV disease according to the WHO clinical classification, since CD4 count is not available in our hospital. Latter on, the patient was started on Nevirapine (NVP), Zidovudine (AZT, ZDV), Lamivudine (3TC), twice per day which led to the improvement of her dysphagia<sup>[9]</sup>.

A repeat endoscopy was done seven months later that showed the complete disappearance of her oesophageal ulcers (see fig.4).

#### **DISCUSSION:**

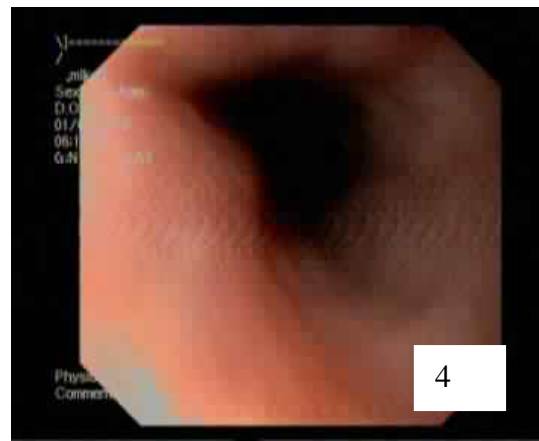
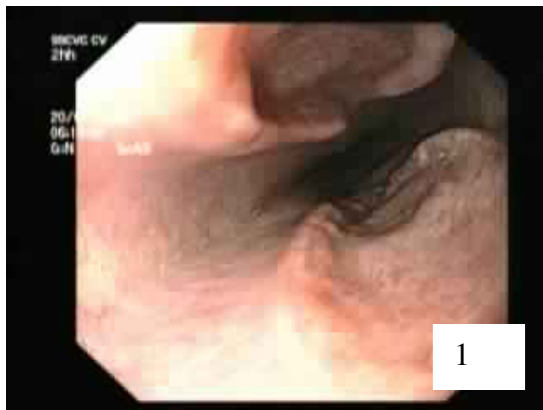
The oesophageal disease is a common complication in patient with human immunodeficiency virus (HIV), it occurs in as many as 30% of these patients

at some time during the infection<sup>[5]</sup>. Oesophageal ulceration is an important cause of morbidity in these patients and may result from many different causes<sup>[5]</sup>. Wilcox and colleagues, in a prospective cohort study of 100 HIV patients with the oesophageal ulcer, showed that 45% of them had cytomegalovirus, 40 had idiopathic ulcers, 5 had herpes simplex virus esophagitis, and 5 had candida oesophagitis<sup>[7]</sup>.

In a cohort of 154 patients, Connolly and colleagues identified 48 patients with oesophageal symptoms, all of whom had an endoscopy. The most frequently identified cause of ulceration was candida oesophagitis, which occurred in 26 patients and with other possible causes in 12 patients. Cytomegalovirus and herpes simplex virus esophagitis have identified the cause in 4 patients; the idiopathic oesophageal ulcer was seen in 3 patients, and one patient has reflux associated ulceration<sup>[5]</sup>. Bonacini and colleagues<sup>[6]</sup>, identified 19 of 110 patients with oesophageal symptoms in whom an erosion or ulcer was endoscopically identified. As in the study by Connolly and colleagues<sup>[5]</sup>, approximately 25% of patients considered by Bonacini and colleagues had at least two causes identified, usually candida ulceration plus another cause. Of the 19 patients with esophageal erosion or ulcer, 11 had a viral infection (7 had cytomegalovirus infection, 3 had herpes simplex virus infection, and 1 had both cytomegalovirus and herpes simplex virus infection). Six patients had no cause identified.

What is interesting about the case we are presenting is that it showed two kissing oesophageal ulcers, with dilated vascular channels, situated intramucosal and in the submucosa. This is more or less resembling angiokeratoma. As Imperial and Helwing discussed in 1967, angiokeratomas are not true angiomas but rather telangiectasias of pre-

existing vessels <sup>[8]</sup>. Angiokeratoma corporae diffusum (Fabry's disease) was carefully excluded in this patient by the presence of a normal alpha-galactosidase A activity in her blood. We postulate that the cause of dysphagia in our patient is that food is trapped between the two kissing oesophageal ulcers. Interestingly, our patient showed a rapid response to HAART, and the oesophageal ulcers have healed completely in a few months' time (see fig.2).



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