

Clinical Challenge 6:

You have been asked to review an 8 month old male infant who is admitted under general paediatric team. He was admitted because of a blistering eruption of palms and soles noticed in the last week which then became widespread in the last couple of days. In the last 24 hours, baby is refusing his feeds and milk and also had few episodes of vomiting which appeared coffee ground in colour. On examination baby appeared pale, but haemodynamically stable. The baby was afebrile throughout the duration of hospital stay and parents confirmed that there was no fever, foreign travel or contact with any unwell persons in the last few months. The skin had multiple erythematous patches and small, fluid-filled blebs on both hands and feet. Blood tests showed haemoglobin 61 with normal platelets, WBC counts and differentials. Clotting screen was normal. Liver function, renal function tests and CRP was normal. Virology results are awaited. The infant was started on intravenous fluids and IV Omeprazole. You have arranged an urgent OGD which showed friable oesophageal mucosa with 2 bloody, fluid-filled blebs ranging from 2 to 4 cm in diameter. More blebs developed on endoscopic contact with the oesophageal mucosa. Biopsies of one of these lesions resulted in “peeling” of the mucosa. The rest of the endoscopic appearance was normal.

What is the diagnosis?

Answer to Clinical Challenge 6

This patient had Oesophageal involvement of **bullous pemphigoid**.

Congratulations to **Dr Protima Amon** for getting the correct answer.

Bullous pemphigoid (BP) is an acquired bullous disorder that is not often seen in children, but it can present with considerable clinical and histologic overlap with other acquired or congenital blistering disorders. This patient’s bullous pemphigoid was classified as type II because of the presence of antibodies against the 180-kDa basement membrane antigen. The patient was started on prednisolone, after consulting dermatology team, with rapid improvement of symptoms. The prednisone was tapered down and at follow-up and he did not have any gastrointestinal or skin problems.

Other diseases presenting with vesico-bullous or exfoliative esophagitis include herpes simplex virus, oesophagitis dissecans superficialis, graft-versus-host disease, reactions to blood transfusion, and amyloidosis- most often seen in adult patients, but can be seen in children as well.