

## Case Report

# Primary duodenal Burkitt lymphoma in a paediatric patient, presenting as a protein-losing enteropathy

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## ABSTRACT

Lymphomas of gastrointestinal system are usually of non-Hodgkin's type. Primary Burkitt lymphoma involving the duodenum is very rare in paediatric population. It may present as protein losing enteropathy in adults which may lead to generalized edema, however in paediatric population these cases are associated with colicky or persistent abdominal pain, weight loss, fever, gut bleeding or obstruction. We report a case of a young male child with primary duodenal Burkitt Lymphoma presenting as protein losing enteropathy.

**KEY WORDS:** Burkitt Lymphoma, Protein losing enteropathy, Primary duodenal.

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## INTRODUCTION

Burkitt's lymphoma (also called small noncleaved cell lymphoma) is a type of non-Hodgkin's lymphoma. It is a malignant proliferation of undifferentiated B-lymphocytes that most often affects children, in all age groups, majority of the patients are male with a 3 or 4:1 male: female ratio.<sup>1</sup> Burkitt lymphoma is a high grade, rapidly growing neoplasm, early diagnosis and treatment offers the best chance of survival. The common gastrointestinal involvement sites include, the duodenum, cecum, ascending colon and jejunum,<sup>2</sup> Because of the paucity of lymphoid tissue in the duodenum, primary duodenal lymphoma is a rare entity, very few cases associated with hypoalbuminemia and oedema have been reported. It accounts for less than 5% of all small bowel lymphomas.<sup>3</sup>

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## CASE REPORT

A five years old boy was admitted with history of fever, generalized body swelling and diarrhea for the last three months, he was passing 6-8 foul smelling stools per day which were loose, bulky and greasy. Patient complained of pain abdomen, off and on vomiting, poor appetite and weight loss during this period. There was no history of oliguria, haematuria, jaundice, bleeding from any site and worms in stool. He appeared pale, grossly oedematous with abdominal ascities. His blood pressure was 85/60mmHg. There was no lymphadenopathy, hepatosplenomegaly or abdominal mass. His weight and height were below third centile. Bone age was 3.5 years. Laboratory investigations revealed hemoglobin. 8.5g/dl, total leukocyte count.8.8x10<sup>9</sup>/l, polys. 58%, lymphos. 36%, monos. 6%, platelets 307x10<sup>9</sup>/l, erythrocyte sedimentation rate 52 mm/1<sup>st</sup> hour, serum sodium 136mmol/l, serum potassium 3.5 mmol/l, serum albumin 1.8 gm/dl, serum calcium 8.7mg/dl, serum ALT 45 IU/l, urea 14 mg/dl, creatinine 0.5 mg/dl, Urine examination was normal with no proteinuria or haematuria. Red blood cells morphology, stool examination, lipid profile, prothrombin time and activated partial thromboplastin time were within normal range. Viral screening for hepatitis B and C were negative. X-Ray Chest was normal except obliteration of right costophrenic angle.

As there was hypoalbuminemia and the patient was grossly oedematous inspite of normal liver functions, renal functions and urinalysis, so he was thought to be a case of protein losing enteropathy and/or malabsorption syndrome. We got his anti tissue transglutaminase IgA done that was within normal range (1.04 U/ml). Ultrasound abdomen revealed gross ascities, thick walled gut especially third and fourth parts of duodenum and proximal jejunum but there was no abdominal lymphadenopathy. On upper gastrointestinal endoscopy there were numerous small tumorous swellings in third and fourth parts of duodenum. Three biopsy samples were taken and sent for histopathology which revealed high grade B-cell non-Hodgkin lymphoma which was diffusely positive for CD 20 in tumour cells representing Burkitt lymphoma. This patient was referred to the Paediatric Oncologist for further evaluation and management.

### DISCUSSION

Burkitt lymphoma (BL) is a highly aggressive B-cell neoplasm that can present in one of three distinct clinical forms: endemic, sporadic, and immunodeficiency-associated. Although they are histologically identical and have similar clinical behavior, there are differences in epidemiological, clinical and genetic features between the three forms.<sup>4</sup> The endemic Burkitt lymphoma refers to those cases occurring in African children (usually 4-7 yrs of age) involving the bones of jaw and other facial bones, kidneys, gastrointestinal tract, ovaries, breast and other extra-nodal sites. Sporadic cases or American variety occurs worldwide. The abdomen especially the ileocecal area is the most common site of involvement. Immunodeficiency associated Burkitt lymphoma mainly occur in the HIV infected patients.

Burkitt lymphoma is a tumour of B-cell origin that can be diagnosed morphologically with a high degree of accuracy. Patients present with peripheral lymphadenopathy or intra abdominal masses. The disease is rapidly progressive, with a propensity for central nervous system metastases. Typical clinical features include colicky or persistent abdominal pain, weight loss, fever and some patients have associated protein losing enteropathy.<sup>5</sup> Rarely patients may present with jaundice due to obstruction of common bile duct.<sup>6</sup> Chieng et al described a case of childhood gastrointestinal tract Burkitt lymphoma with protein losing enteropathy<sup>7</sup>, more recently, Shital et al also reported a case of paediatric Burkitt lymphoma with duodenum as a primary site.<sup>8</sup>

Studies in Paediatric populations have shown that CT scan is an important mode of investigation in characterizing the morphological pattern. Similar findings can be seen on MRI studies in patients with small intestinal tumour, whereas ultrasonography shows small submucosal nodules which may be overlooked.<sup>9,10</sup>

Our patient was of primary duodenal Burkitt lymphoma in childhood which presented with protein losing enteropathy in which initial investigations were non diagnostic. However upper gastrointestinal endoscopic duodenal biopsy revealed involvement of duodenal mucosa with primary Burkitt lymphoma hence he was later referred to the Paediatric Oncology Department for further evaluation and management.

### CONCLUSION

Paediatric duodenal Burkitt lymphoma is a rare entity which may present unusually with protein losing enteropathy and gross edema. Endoscopic duodenal biopsy may be a useful diagnostic tool. The clinical outcome may be excellent if diagnosis is made earlier, even when presenting with rare clinical manifestations.

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### Authors Contribution:

**Rabbani MW, Aziz MT:** Conceived, designed and manuscript writing.

**Ali Z, Khan WI, Aslam M:** Editing and review of manuscript.