

Primary Pancreatic Burkitt's Lymphoma Presenting as Obstructive Jaundice: A Case Report

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I. Introduction

Non Hodgkin's Lymphoma (NHL) is known to disseminate into extra nodal sites constituting 20-30% of all the lymphomas, with the stomach, skin, oral cavity and pharynx, small intestine, and central nervous system being the common sites⁴. Primary Pancreatic Lymphoma (PPL) accounts for about 0.2-2% of the NHLs and among these Burkitt's Lymphoma is uncommon.

The diagnostic criteria of PPL includes Mass predominantly located within the pancreas, with grossly involved lymph nodes confined to the peripancreatic region, no palpable superficial lymphadenopathy, no hepatic or splenic involvement, no mediastinal nodal enlargement on chest radiography and normal white blood cell count⁵. Unlike pancreatic adenocarcinoma for which surgery is the mainstay of treatment, chemotherapy has been shown to be the most efficacious first line of treatment for PPL. Thereby necessitating its importance in evaluation due to its high cure rate.

Here we have a case of young adult male presenting with symptoms of obstructive jaundice and on detailed evaluation was diagnosed with pancreatic ampullary Burkitt's Lymphoma.

II. Case report

31year old male with no significant past medical history who presented with epigastric pain since 6 months, associated progressive yellowish discolouration of skin and sclera passage of pale coloured stools, high coloured urine with anorexia. On clinical examination, his vitals were stable and systemic examination revealed tenderness in the epigastric region.

He was evaluated elsewhere and CT and MRI was done suggestive of diffusely enlarged pancreas with multiple varying sized mass lesions scattered all over the pancreas, one of the mass in the uncinate process caused CBD obstruction with IHBRD. His haematological analysis showed a total leukocyte count of 5900cells/cu.mm and differential count was at Neutrophils-77% Lymphocytes-13% Monocytes-7% Eosinophils-3%, a deranged liver function with Total Bilirubin 13.8mg/dL and Direct Bilirubin of 12.05mg/dL, Alkaline Phosphatase:637IU/L, Aspartate Transaminase (AST): 422IU/L, Alanine Transaminase (ALP):644IU/L.

Endoscopic ultrasound guided fine needle aspiration of the pancreatic mass was done with ampullary biopsy for IHC. Meanwhile the patient was subjected to ERCP + stenting to relieve his obstructive symptoms. ERCP showed distal CBD obstruction due to mass in the head of the pancreas and was stented. Histopathological examination revealed features of Non Hodgkin's Lymphoma while immuno-histochemistry markers were positive for CD-3,5,10,20 and cMYC with Ki67 of 90-95% proving Burkitt's Lymphoma.

PET CT was done and suggested active disease in the diffusely bulky pancreas with necrotic areas and hypermetabolic areas in the small bowel loops, upper pole of left kidney, left testis, level II, precarinal and internal mammary nodes. He was referred to medical oncology and has been started on chemotherapy.

III. Discussion

PPLs are extremely rare, comprising less than 2% of extra nodal malignant lymphomas and 0.5% of pancreatic tumors¹, although nearly one-third of non-Hodgkin's lymphoma patients will ultimately develop some form of pancreatic involvement². Diffuse large-cell lymphomas comprise the bulk of PPLs while Burkitt's lymphoma, is exceptionally rare.

Burkitt's lymphoma is attributed to translocations between MYC gene on chromosome 8 and the IgH gene on chromosome 14, while MYC-*kappa* light chain and MYC-*lambda* light chain translocations are also described³

Unlike pancreatic adenocarcinoma, which is treated surgically, chemotherapy has been shown to be the most efficacious first-line treatment for PPL with 72% of the patient showing no disease at the end of 36 months.³¹ using regimes like CVP, MACOP-B, and CHOP. The further addition of rituximab, a monoclonal antibody targeting the CD20 B-cell antigen, to the CHOP regimen (R-CHOP) has proved to be more effective

Multiple chemotherapeutic agents have proven to be a curative in Burkitt's Lymphoma, but an optimal initial therapeutic approach to the disease has not yet been defined due to lack of any supportive trials towards the treatment; adding on is the rarity of the disease.

IV. Conclusion

PPLs are a class of rare cancers affecting the pancreas, of which Burkitt's lymphoma comprises an exceptionally rare subset. However, as efficacious treatments for the disease exist and differ markedly from that of pancreatic adenocarcinoma, it is important that PPLs more generally and Burkitt's lymphoma in particular be included in the differential when evaluating a probable pancreatic neoplasm.

References

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