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Isolated Non-Hodgkin's Lymphoma of the Pancreas: Case Report and Review of Literature

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1. Introduction

Extranodal non-Hodgkin's lymphomas (NHLs) represent up to 30-40% of all NHL cases. The gastrointestinal tract is the most commonly involved extranodal site; accounting for about half of such cases [1]. non-Hodgkin's lymphomas often invade extra-lymphatic organs. Most primary pancreatic lymphomas (PPL) are non-Hodgkin's Lymphomas. More than 25 percent of non-Hodgkin's lymphomas originate from extra-lymphatic organs, about 30 percent of which may involve the pancreas(2). Primary pancreatic lymphoma (PPL) is rare, comprising less than 0.5% of pancreatic tumours[3]. Have suggested a strong male predominance (maleto- female ratio of 7:1). The patients range in age from 35 to 75 years (mean age: 55 years) [4].

Presentation of primary pancreatic lymphoma[5]

Symptom/sign	Incidence
Abdominal pain	83%
abdominal mass	58%
Weight loss	50%
Jaundice	37%
Nausea	34%
Vomiting	18%
Diarrohea	12%
Pancreatitis	12%
Bowel obstruction	12%
Diarrohea	12%
Fatigue	9%
Fever, chills, night sweats	2-7%
Gastrointestinal bleeding	2%
Gastric outlet obstruction	2%

Etiology of ppl is generally unknown. Behrns' clinical and diagnostic criteria of PPL include: mass predominantly 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 radiograph, and normal white cell count [6]. Percutaneous ultrasound (US), endoscopic ultrasound (EUS) and computed tomography (CT) scan are well-established procedures to evaluate pancreatic masses. Cytohistological diagnosis is mandatory for diagnosis. IHC also help in confirming diagnosis.

2. Case Report

65 year old male patient presented with a c/o pain right side of abdomen, loss of appetite. CECT abdomed showed grossly enlarged body and tail of pancreas, homogenous moderately enhancement. Mass with retro peritoneal lymphadenopathy. His TLC count was raised. EUS was done and FNAC from pancreatic region was taken which was suggestive of NHL. Immunophenotyping revealed absolute CD20 positive cell count. IHC cells stained positive foe CD20 (B cell marker). CT chest & bone marrow was done to rule out infiltration. So a diagnosis of primary pancreatic lymphoma (NHL) was made. Patient received 1 cycle R-CHOP. Later he presented in emergency Geetanjali medical college in comatose state. He was managed for septicemia, electrolyte imbalance , generalized weakness for one month in ICU. Upon recovery patient wasplanned for 6 cycles R-CHOP. Due to presence of retroperitoneal lymphadenopathy (in pretreatment CT& post chemotherapy) IFRT to para aortic lymph nodes was planned and delivered. Total dose of 36Gy was delivered in 20#, 1.8 Gy/day(VMAT-IGRT). His post treatment PET CT revealed no recurrent or residual mass lesion. His follow PET CT showed no residual/ recurrent mass lesion in pancreas, metabolically inactive submandibular, para aortic, precarinal , subcarinal, hilar, retroperitoneal lymphnodes. his latest CT abdomen revealed no abnormality. He is disease and symptom free for 21 months.

3. Discussion

PPL can be difficult to differentiate from pancreatic adenocarcinoma without definitive pathological diagnosis. Reliance on symptoms, imaging and tumour markers – in the absence of definitive pathological diagnosis of suspected pancreatic adenocarcinoma – can potentially result in the misdiagnosis of a small minority of potentially curable patients.

Treatment options[5]

Modality	indication	Comment
Chemotherapy	Primary treatment after tissue diagnosis	Primary treatment
radiotherapy	Adjunct to chemotherapy	Adjuvant treatment
surgery		No role

Arcariet al. showed outcome of 5-8 months with Surgery plus chemotherapy & 69-80 months with Chemotherapy[7].

Bouvet et al reported ten in eleven PPL patients, have been performed explorative surgery, only in three of them the tumor could be fully resected. Among eight unresected cases, seven cases were treated with combined CHOP chemotherapy and radiotherapy with dosage of 30 to 45 Gy. The median survival time was 67 mo (11-191 mo)[8].

Behrnset al. reported outcome of 13 months with chemotherapy alone, 22 months with external radiotherapy, 36 months with Chemotherapy plus external radiation therapy[6].

Therefore, the first choice for PPL treatment should be combination of chemotherapy and radiotherapy,rather than surgery. With the advancement of techniques, surgery seems to be only effective when FNA is not available or diagnosis can not be made on histology. It has already been proved that single pancreas resection could not improve the survival rate of PPL but cause more complications.

In conclusion total pancreatectomy is considered to have no impact on survival and with its associatedmorbidities, is not generally recommended for diagnosis and treatment of PPL. As a result, PPL will not be such a disease with poor prognosis in the future.

4. Conclusion

An exceedingly rare entity, isolated PPLs need to be differentiated from pancreatic adenocarcinomas since management is on the lines of other extralymphatic lymphomas and prognosis is significantly better.

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