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RESEARCH ARTICLE

CHOLESTATIC JAUNDICE REVEALING A PANCREATIC BURKITT LYMPHOMA IN A CHILD: A CASE REPORT

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Abstract

Burkitt lymphoma is an aggressive form of non-Hodgkin lymphoma, characterized by rapid proliferation of B cells. It typically presents with abdominal masses or extra-abdominal involvement. Although rare, it can be a potential cause of cholestatic jaundice due to extrinsic compression of the bile ducts or direct infiltration of the liver or pancreas. In children, cholestatic jaundice can be associated with various etiologies, including infections, congenital liver diseases, metabolic syndromes, and tumors. The discovery of a rare and malignant cause, such as Burkitt lymphoma, adds complexity to diagnosis and management. In this article, we report a case of a child with cholestatic jaundice as a presenting symptom of pancreatic Burkitt lymphoma. We discuss clinical aspects, diagnostic modalities, and therapeutic options, highlighting the challenges encountered in diagnosing and managing this atypical presentation. A review of the literature shows that cases of Burkitt lymphoma with pancreatic involvement and cholestatic presentation are rarely documented, making this case particularly noteworthy.

Introduction:-

Burkitt lymphoma is a highly aggressive non-Hodgkin B-cell lymphoma, frequently observed in children and young adults. It typically presents with abdominal masses, jaw involvement, and rarely with pancreatic involvement [1-3] . The clinical presentation of pancreatic Burkitt lymphoma can be deceptive, sometimes manifesting as non-specific symptoms such as cholestatic jaundice, which may delay diagnosis. [2]

The objective of this article is to describe the case of a child with cholestatic jaundice revealing pancreatic Burkitt lymphoma, to discuss the diagnostic methods and therapeutic strategies implemented. We aim to raise awareness among clinicians about this atypical presentation to promote early recognition and appropriate management.

Case presentation:

We report the case of a 15-year-old patient who presented to the pediatric emergency department with rapidly progressive symptoms that had developed over the past month and a half, including epigastric pain and cholestatic jaundice in the context of general deterioration (weight loss of 6 kg per month; fatigue; anorexia). Clinical examination revealed a stable patient neurologically, hemodynamically, and respiratorily, with generalized cholestatic jaundice, a distended and tense abdomen, painful on palpation, and signs of portal hypertension:

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collateral venous circulation with shifting dullness in the flanks, palpable lymphadenopathy in the axillary and inguinal regions bilaterally (with the largest measuring 2 cm), and edema of the lower limbs. The rest of the physical examination was unremarkable.

Abdominal CT scan revealed a tumor process centered on the head and isthmus of the pancreas, locally advanced (Figure 1), associated with mesenteric lymphadenopathy, moderate ascites, and portal vein thrombosis. The child underwent an endoscopic ultrasound-guided biopsy, and histological analysis confirmed Burkitt lymphoma (Figure 2). Initial tumor lysis syndrome was disrupted, and the patient was placed on hyperhydration, alkaline treatment, and allopurinol with strict biological monitoring. The staging workup (Body scan, bone marrow biopsy, testicular ultrasound, and cerebrospinal fluid analysis) was unremarkable, as was the pre-treatment assessment (echocardiogram and serologies for HIV, HCV, HBV). LDH was elevated at 1170 IU, lipase was high at 110 IU, with hepatic cytolysis and cholestasis. The patient was started on chemotherapy, treated as Group B. Follow-up imaging showed an estimated 50% regression of the pancreatic tumor. The patient developed severe bone marrow aplasia at the end of the 5th cycle of chemotherapy, with severe neutropenia leading to death from septic shock.

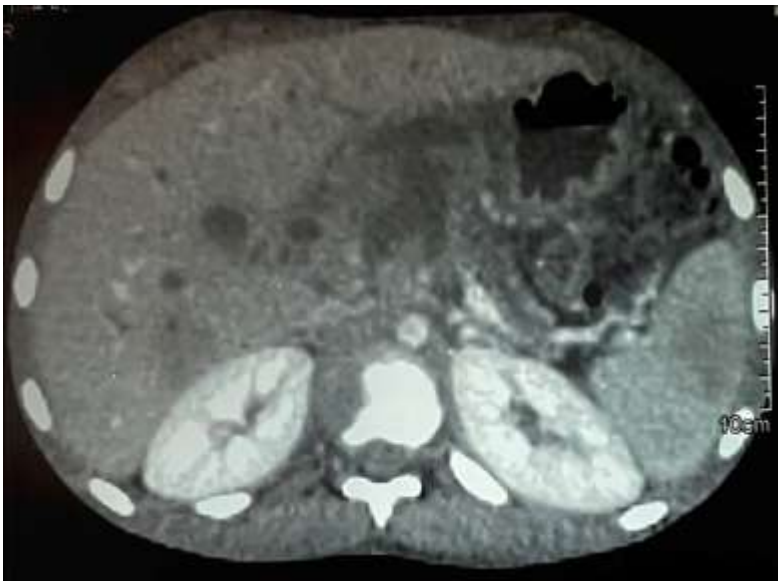

Figure 1:- Abdominal CT scan revealing a tumor process centered on the head and isthmus of the pancreas, locally advanced.

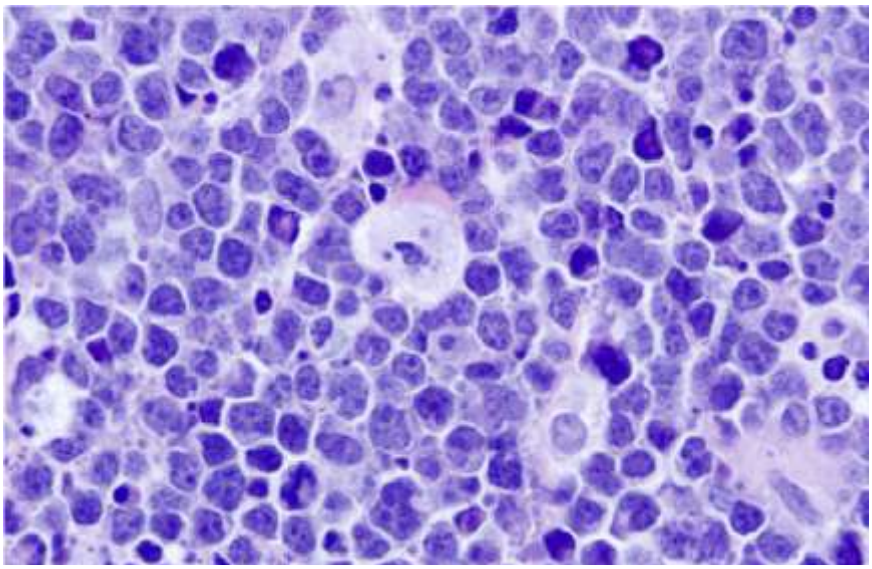

Figure 2:- Histopathological features of Burkitt lymphoma under the microscope.

Discussion:-

Burkitt lymphoma is a malignant tumor proliferation of B-lymphoid cells, characterized by a translocation and dysregulation of the c-myc gene on chromosome 8. It is a high-grade, aggressive non-Hodgkin lymphoma with significant tumor dissemination, particularly to the bone marrow and central nervous system [4] . There are three distinct clinical forms of Burkitt lymphoma: endemic, sporadic, and associated with immunodeficiency [5].

The endemic form, known as African Burkitt lymphoma, is characterized by mandibular involvement and its association with Epstein-Barr virus (EBV), making it a model of virus-associated tumors. Although the exact role of EBV in the genesis of this lymphoma is still not fully understood, it is notable for its geographical distribution in tropical countries [5-6] . The immunodeficiency-associated Burkitt lymphoma is generally associated with HIV infection. The sporadic form, also known as American Burkitt lymphoma, is predominantly abdominal in 70-90% of cases, with less frequent maxillary involvement (10-15%).[7-8]

In Morocco, the incidence of Burkitt lymphoma is unknown. Its epidemiological profile would likely correspond more to the sporadic type. In a study by Madani et al. in 2005, maxillary involvement was found in 9.5% of cases, with abdominal involvement in 73.5%. Otmani et al. in 2008 found that among 452 cases of non-Hodgkin lymphoma, 8% had oral involvement with Burkitt lymphoma [9].

The ileocecal region is the most common site for sporadic lymphoma. It usually presents as a palpable mass and ascites, involving the distal ileum, stomach, cecum, and/or mesentery. Initial symptoms may include those related to intestinal obstruction or gastrointestinal bleeding, often mimicking acute appendicitis or intestinal intussusception. Pancreatic involvement in Burkitt lymphoma is rare[10]. Primary pancreatic lymphoma is much less common than secondary involvement, occurring predominantly in individuals over 60 years old, which was not the case in our observation [11]. . The clinical symptoms are non-specific and mainly include: epigastric pain (83%), abdominal mass (58%), weight loss (50%), and jaundice (42%). Our patient presented with jaundice, epigastric pain, and general deterioration [12].

Regarding complementary examinations: no specific biological marker is available, though lactate dehydrogenase may be elevated. Radiological exams help guide the diagnosis: CT scans, MRI, and PET scans. The diagnosis is confirmed by histopathological analysis of a percutaneous pancreatic biopsy guided by ultrasound or CT scan, or an endoscopic ultrasound-guided biopsy, which is the reference examination and confirmed the diagnosis in our patient. Tumor lysis syndrome should be regularly monitored due to the high risk. The staging workup includes cervico-thoraco-abdomino-pelvic CT, bone marrow biopsy, testicular ultrasound, and cerebrospinal fluid analysis [12].

Burkitt lymphoma is one of the fastest-growing pediatric tumors, with a doubling time of about 24 hours, which underscores the importance of rapid diagnosis for initiating appropriate treatment. Polychemotherapy is currently the mainstay of treatment due to the tumor's high chemosensitivity. The prognosis depends on the extent of initial disease and the speed of treatment initiation [13]..

Treatment for Burkitt lymphoma focuses on the prevention and treatment of tumor lysis syndrome and intensive polychemotherapy. The most effective agents include: cyclophosphamide, methotrexate, cytarabine, vincristine, and doxorubicin [13]..

A literature review shows that cases of Burkitt lymphoma with pancreatic involvement and cholestatic presentation are rarely documented. Amodio et al. report the case of a six-year-old child with primary pancreatic Burkitt lymphoma, initially presenting with cholestatic jaundice. This case highlighted the importance of imaging, particularly MRI, in this pathology and the necessity of early and accurate diagnosis through endoscopic biopsy to differentiate Burkitt lymphoma from other pancreatic or ampullary tumors. The child responded well to chemotherapy [14]..

Although cholestatic jaundice is a common symptom in pancreatic Burkitt lymphoma, its absence does not necessarily exclude the diagnosis. H. Kim et al. report the case of a 16-year-old adolescent with pancreatic Burkitt lymphoma, which was revealed by severe abdominal pain mimicking acute pancreatitis. The patient was treated with a combined chemotherapy regimen, leading to significant improvement in pain and tumor reduction [15].

These cases underscore the rarity of pancreatic Burkitt lymphoma and the critical importance of a multidisciplinary approach involving imaging, biopsy, and appropriate chemotherapy to achieve favorable outcomes.

Conclusion:-

In conclusion, this case illustrates the complexity and diversity of clinical presentations of Burkitt lymphoma in children and highlights the importance of a rigorous diagnostic approach and multidisciplinary collaboration in managing unexplained cholestatic jaundice..

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