Report Of A Case Of Abdominal Burkitt's Lymphoma Presenting As Localised Right Iliac Fossa Pain Mimicking Acute Appendicitis

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Citation

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Abstract

Burkitt's lymphoma is a high grade B-cell neoplasm and has two major forms; the endemic(African) and non endemic (sporadic)form. It is one of the fastest growing malignancies in human, with a very high growth fraction. The African form often involves the maxilla or mandible. In contrast the sporadic form usually involves abdominal organs with the most common involvement of the distal ileum, caecum or mesentery. The Epstein-Barr Virus(EBV) has been implicated strongly in the African form while such relationship is less clear (approx 20%)in the sporadic form.(1),

The sporadic form commonly present with abdominal swelling ,pain or fullness. Some patients present with symptoms of bowel obstruction secondary to ileo-caecal intussusception caused by tumor growth. We present a case report of a 14 year old boy who presented with acute localized right iliac fossa pain of few hours duration with associated nausea and vomiting mimicking acute appendicitis. He was healthy in the past without any complaint. Such localized RIF presentations are rare. This case also emphasis the importance of medical diagnosis either by imaging (USG/CT) or direct visualization (laparoscopy) to characterize appendicular lesions and to select the appropriate surgical procedure.(2)

CASE REPORT

A 14year old boy was admitted from casualty department with complaints of acute right iliac fossa pain. The pain was of few hours duration, constant without any waxing or waning, moderate to severe in intensity, with no radiation and associated with nausea and a couple of bouts of vomiting. He had no urinary or bowel symptoms.

CLINICAL EXAMINATION

On examination he was tender in the right iliac fossa region but generally the abdomen was soft with no rigidity ,guarding or rebound tenderness. Bowel sounds were audible normally with no hyperkinetic rushes or metallic tinkling. General physical examinations revealed moderately enlarged bilateral cervical lymphadenopathy. The palatine tonsils were enlarged and engorged and signs of pharyngitis were present on oral examination. He had a few crepitations in the right mid zone but apart from this there was no abnormal finding. His temperature was raised at 37.8 C. Routine urinalysis showed moderate amount of ketone and trace of protein.

In the past the patient had not had any serious illness. He has been fully vaccinated. He has 3 siblings aged 10,14 and 16 years-all of whom are in good health.

A provisional diagnosis of acute appendicitis or mesenteric adenitis was made. A diagnostic laparoscopy was performed which revealed the appendix to be normal but a mass in the small bowel. A formal laparatomy with a midline incision was performed. A tumor was found in the ileum approx 12 inches from the ileo-caecal valve. A large mass was found in the mesentery and a further mass in the mesentery of the ascending colon. In addition there were multiple enlarged mesenteric nodes. A right hemicolectomy was performed removing all 3 tumors and an end to side ileo-tranverse anastomosis was done.

INVESTIGATIONS

MACROSCOPICAL

The preliminary macroscopic pathological examination of terminal ileum revealed the mucosa to be grossly altered with multiple thickened polyps which were sessile. The mass appeared to stretch the serosa overlying the bowel wall but

did not appear to penetrate it and extended only focally into the mesentery. The area of the mass showed an irregular, vaguely polypoid mucosal lesion measuring 3cm maximum in dimension occupying 80% of the bowel wall circumference at that region. The bowel mucosa between the tumor mass and the resection margin showed a distinct micronodularity, possibly lymphoid hyperplasia. This was much less marked on the other side of the tumor and it disappeared altogether 10 cm beyond the tumor with apparently normal mucosa forming the final 10-12 cm of bowel. The caecum and the ascending colon showed no mucosal changes.

There were lymph nodes approx 5-4-4 cm in dimension consisting of solid creamy white tissue. Smear taken from this revealed highly cellular lymphoma with blast cells and intervening macrophages suggestive of the appearance of undifferentiated lymphoma of Burkitt's type..

MICROSCOPICAL

Microcopic examination showed that the small bowel was extensively infiltrated by a cellular neoplasm with a diffuse growth pattern. The tumor extended from the mucosa through the full thickness of the bowel wall and was focally identified on the serosa. The mucosa overlying the neoplasm was attenuated and focally ulcerated. The tumor was composed of cells of a high nuclear: cytoplasmic ratio and the nuclear appearances varied dramatically depending on the quality of fixation. In areas they appeared intermediate to large size with vesicular nuclei and multiple small nucleoli. Elsewhere the tumor cells were smaller and bluer in appearance. In all locations there was a brisk mitotic and apoptotic rate with substantial population of tangible body macrophages. Eosinophils were also identified mixed in with the malignant cell population. The tumor showed extensive areas of necrosis, with the mesenteric nodule in particular, large necrotic. Even walls within the bowel wall showed marked degenerative changes with a pseudo alveolar pattern. The bowel away from the tumor showed no enteropathic changes and the nodularity identified grossly correspond to the profound lymphoid hyperplasia with a normal lymphoid tissue microarchitecture..

IMMUNOLOGICAL STUDY

Immunohistochemical status showed the malignant cell population was positive for lymphoid markers CD45, CD20 and CD79a but negative for T cell markers CD3.The histological features were those of high grade diffuse non Hodgkin lymphoma of B cell origin and compatible with Burkitts' lymphoma.

ABDOMINAL USG

An abdominal USG showed slight prominence of the pelvis of each kidney but no evidence of calyceal dilatation.The appearance were thought to be probably due to prominent extrarenal pelvis.The liver,spleen,pancreas,gallbladder and aorta appeared normal.No paraaortic lymphadenopathy was identified.

HEMATOLOGY & BIOCHEMISTRY

The full blood count (FBC) was normal. Urea and Electrolytes(U/E) showed hypochloraemia, hypocalcaemia, hypomagnesaemia, hypoproteinaemia, and hypoalbuminaemia. His AST was moderately elevated at 60. The LDH was 366. Plasma urate was normal. Serum immunoglobulins did not identify an underlying immune deficiency. His CSF did not show any evidence of lymphoma infiltration.

BONE MARROW EXAMINATION

Bilateral iliac crest trephine biopsies showed no bone marrow involvement .by tumor. Given the above findings- the tumor was group B.

TREATMENT

Following discussion with the patient and his parents, the nature of the malignancy was explained. We outlined a probable good outline and commenced treatment using triple agent mild chemotherapy couple of days following surgery. On that day he received intrathecal Methotrexate and hydrocortisone . This treatment was followed one week later by a much more intensive programme which utilized Vincristine, Prednisolone, Methotrexate, Cyclophosphamide, Doxorubicin and further intrathecal chemotherapy.

The prognosis for this particular tumor has improved drastically over the years since the adoption of the very intensive chemotherapy approach. Survival is expected in the region of 80%.Although the patient was initially relatively uncomfortable as he was still recovering from the surgery .On his next admission a month later the patient seemed comfortable-his health and quality of life were excellent. The current study involves a number of randomizations. This patient was randomized to ARM B1 of the FAB LMB Protocol which involved treatment lasting 3 to 4 months. Radiation therapy should not be required assuming that there is no evidence of tumor recurrence.

His yearly checkup seemed satisfactory.

DISCUSSION

The advent of immunohistological and molecular technique have enabled the comprehensive characterization of many lymphoma entities. The majority of primary intestinal lymphomas are of B- cell lineage and most of these are high grade tumors. By morphology they may be classified as diffuse large B-cell lymphomas of centroblastic, immunoblastic or plasmablastic type and Burkitt's lymphoma.

Burkitt's lymphoma occur predominantly in the terminal ileum and affect children or young adults as in this particular case and might present as abdominal pain or fullness([3]).The diagnostic category of Burkitt's lymphoma encompasses a closely related group of aggressive B-cell tumors that includes sporadic, endemic and HIV associated subtypes . All Subtypes are characterized by chromosomal rearrangements involving the c-myc proto-oncogenes that lead to its inappropriate expression.([4]) In Burkitt's there is a characteristic cytogenetic abnormality of translocation between the long arms of chromosome 8 and 14. The proto-oncogenes c-myc is translocated from its normal position on chromosome 8 to the heavy chain locus on chromosome 14. Cells commited to B-cells differentiation are likely to have enhanced expression of this heavy chain locus and it is likely that overexpression of c-myc (in its new anomalous position) is related to malignant transformation.([5])

For patients with limited diseases, including localized extra abdominal or completely resected abdominal disease (NCI Stages A and AR)a long term survival rate>90% can be achieved with combination chemotherapy([6,7,8,9,10,11,12,13]). Treatment options include:

COMP- Cyclophosphamide+Vincristine(Oncovin)+ Methotrexate+Prednisone.
CHOP-plus methotrexate
NHL-BFM90-
Prednisone+dexamethasone+vincristine+doxorubicin+cyclophosphamide+ifosfamide+etoposide+cytarabine+methotrexate
French LMB-89-high dose cyclophosphamide + high dose methotrexate/leucovorin+cytarabine+vincristine+prednisone +doxorubicin
CCG-5961- Reduction in intensification of the French LMB-89 regimen

Link et al ([14,15]) demonstrated that a 9 week regimen of cyclophosphamide, doxorubicin, vincristine and prednisone plus intrathecal prophylaxis was as effective for early stage disease as a longer regimen that included a continuation phase with or without radiation. With this and similar regimens for limited disease, cure rates exceed 90%with minimal toxicity. Thus, less intensive therapy is under evaluation as a potentially effective way to avoid unnecessary toxicity for patients with early stage disease, while achieving similar cure rates obtained with more prolonged treatment.

For patients with extensive disease, a long term survival rate of 70-80% now can be achieved with intensive chemotherapy regimens.([13,16,17,18]) Current treatment options include:

French LMB-89,Total B-St.Jude's:NHL-BFM90NCL-89-C-0041F

Surgical care is employed only for patients with small, completely respectable abdominal tumors or patients with obstruction who cannot begin chemotherapy immediately.

Prognosis in children correlates with the bulk of disease at the time of diagnosis. With appropriate management of the metabolic consequences of rapid cell turnover and with combination chemotherapy and CNS prophylaxis, the survival rate has been improved significantly. Patients with limited (A,AR) disease have an excellent prognosis with a survival rate greater than 90%.Adults with Burkitt's lymphoma, particularly those with advanced stage disease, do more poorly than children with the disease.

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