Different Surgical Modalities in Management of Paediatric Abdominal Lymphoma

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Abstract

Background

The abdomen is one of the most frequent sites for lymphoma in children. The role of surgery has been limited to intra-abdominal resectable tumours or as a diagnostic procedure in case of disseminated disease. Laparotomy without total excision of the tumour does not improve survival; moreover, it may cause complications and delays initiation of chemotherapy.

Aim of the work

This study was undertaken to assess the role of surgery in the management of children and adolescents presenting with intra-abdominal lymphoma in order to create certain criteria to select the proper surgical modality for managing those patients.

Patients and Methods

This case-series, retrospective study was done on 33 patients of abdominal lymphoma over a period of seven years from 2000 to 2007. Patients’ files were reviewed regarding the full clinical examinations, laboratory and radiological investigations as well as surgical and diagnostic procedures. Collected data were tabulated and statistically analyzed using SPSS program package.

Introduction

Pediatric lymphomas are the third most common malignancy in children and accounts for 13% of all childhood cancers. Its incidence increases with increasing age in children1. Hodgkin (HL) and non-Hodgkin’s lymphomas (NHL) constitute 10-15% of total cancer diagnoses in children in the more developed countries, after acute leukemia’s and brain tumours2-4.

Results

Eleven patients (33.3%) presented with huge pelvi-abdominal mass and eleven (33.3%) had generalized lymphadenopathy beside their abdominal affection. The remaining 11 (33.3%) patients presented with symptoms of an acute abdomen. A total of 15 laparotomies were done. 11 patients underwent emergency laparotomy for acute abdomen and 4 patients had elective abdominal exploration. Lymph node biopsies were taken in 7 patients and laparoscopy procedures were performed in 3 patients as a diagnostic tool. Out of the total 33, the remaining 8 patients underwent true cut needle biopsy for diagnosis of their disease.

Conclusion

Surgery still has a role in treatment of lymphoma whether non Hodgkin or Hodgkin’s. However, in disseminated metastatic disease, aggressive debulking of the tumour should be avoided as chemotherapy is to be instituted primarily. Surgical resection does not cause significant change in morbidity or mortality.

Keywords

Abdominal lymphoma in paediatrics, role of surgery
Approximately 60% of paediatric lymphomas are NHL, with the remainder being Hodgkin’s lymphomas (HL). According to the latest classification (2008) with the use of additional immunological and molecular markers, most paediatric NHL can be grouped into four major histological subtypes, a) Burkitt lymphoma, b) diffuse large B-cell lymphoma, c) anaplastic large cell lymphoma and d) lymphoblastic lymphoma. The most important subtype of HL is nodular sclerosis.

Paediatric patients typically present with Burkitt lymphoma with extranodal involvement, specifically occurring in the abdomen in approximately 31% of the cases. The appropriate role of surgery in intra-abdominal Burkitt’s lymphoma although a controversial subject became more defined over the last two decades. Children with localized bowel tumour scan undergo gross total resection of the primary tumour, with low operational morbidity rate.

Patients with abdominal NHL can be divided into two surgical groups; in the first group the tumor is localized anatomically within the abdomen, in this case, the tumor often involves the bowel wall and many of those children present with acute abdominal symptoms suggesting appendicitis or intussusceptions. The majority can undergo complete gross tumor resection, often with a simple bowel resection and re-anastomosis. In the second group; there is extensive intra-abdominal tumor, and presentation with an abdominal mass without acute symptoms is more likely. The mesenteric root and retro peritoneum are heavily involved and attempts at complete excision are associated with a higher complication rate where surgery is on elective basis is for debulking.

A recent study has stated that laparotomy should be limited to cases presenting with acute abdomen and limited resectable disease. In cases of disseminated disease, bone marrow aspiration, cytological investigation of the ascite/pleural effusion, or ultrasound-guided-true-cut needle biopsy of the abdominal mass were a better alternative, had less morbidity than laparotomy and allowed early initiation of chemotherapy.

This study was undertaken to assess the role of surgery in the management of children and adolescents presenting with intra-abdominal lymphomas in our experience and to report our strategy management of abdominal lymphoma in order to reach certain criteria for selecting the proper surgical modality.

**Patients and methods**

A case-series, descriptive study was done on 33 patients of abdominal lymphoma who were treated over the period of 7 years from 2000 to 2007 in Suez canal university hospital, Ismailia governorate and Damanhour medical national institute, Al-Buhayrah governorate, Egypt and the Eastern Province of Saudi Arabia.

Inclusion criteria were children and adolescents aged 2-18 years, both males and females who were suffering of generalized lymphadenopathy and got tissue biopsy revealing lymphoma and evidence of intra-abdominal involvement. Also, included were children and adolescents of similar age and sex who revealed intra-abdominal mass with tissue biopsy revealing lymphoma.

Excluded of this study were patients out of the specified age range, or those without intra-abdominal involvement. Those who had generalized lymphadenopathy or intra-abdominal masses proven not to be lymphoma by tissue biopsy were excluded as well as patients with post-transplantation lymphoma or severe immune-deficiency status.

After approval of the ethical committee; patients’ files were reviewed regarding demographic data, history taking, full clinical examinations including general examination for the vital signs at presentation. Also reviewed were clinical features at presentation such as abdominal pain, abdominal mass and generalized lymphadenopathy. Records of systemic clinical examination including thorough lymph node group’s examination, abdominal examination, chest and cardiac examination were reviewed. Surgical techniques were thoroughly studied by reviewing all the operative details and findings. Post-operative sequence was also recorded in all patients with the final post-operative results.
Results

The age ranged between 2.5 -16 years with a mean of 6.7 years; there were 18 females and 15 males with F/M ratio of 1.2:1. The sociodemographic characteristics of the patients are shown in Table 1.

Eleven patients presented with abdominal mass and eleven had generalized lymphadenopathy beside their abdominal affection which was discovered after investigations. The remaining 11 patients presented with symptoms of an acute abdomen requiring emergent operation; 4 patients with complete intestinal obstruction secondary to tumour matting, 5 with irreducible intussusceptions with tumour acting as the lead point, and 2 patients with suspected appendicitis (Table 2).

The operative records showed a total of 15 laparotomies. Out of them, 11 patients underwent emergency laparotomy for acute abdomen and 4 patients had elective abdominal exploration (EAE). Lymph node biopsies were taken in 7 patients. Moreover, laparoscopy procedures were performed in 3 patients as a diagnostic tool (Table 3). Out of the total 33, the remaining 8 patients underwent true cut needle biopsy for diagnosis of their disease.

<table>
<thead>
<tr>
<th>Diagnostic Method</th>
<th>Number of patients (n=33)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lymph node biopsy</td>
<td>7</td>
</tr>
<tr>
<td>Urgent laparoscopy</td>
<td>3</td>
</tr>
<tr>
<td>Elective exploration</td>
<td>4</td>
</tr>
<tr>
<td>Urgent exploration</td>
<td>11</td>
</tr>
<tr>
<td>True cut needle biopsy</td>
<td>8</td>
</tr>
</tbody>
</table>

Table 3: Diagnostic method and surgical interventions in studied patients

Laparotomy was performed in 15 patients, of the 4 patients with elective abdominal exploration, total tumour resection and small bowel resection anatomies were done in 1 of them, 2 patients were splenectomised and paraaortic liver biopsy while incisional biopsy was performed in 1 patient. In the other 11 patients where urgent abdominal exploration was performed, 5 patients had intussusceptions. Three of them had ileocolic intussusceptions. Resection anastomosis with lymph node biopsy was done in first patient, debulking in the second patient and right hemicolecotomy with ileotransverse anastomosis due to caecal perforation in the third patient. The remaining 2 patients had ileoileal intussusception with lymphadenopathy where reduction and lymph node biopsy was done for one of them and resection anastomosis and lymph node biopsy was done for the other.

In 4 patients, laparotomy revealed intestinal obstruction, in two of them a mesenteric mass was found with perforation, where debulking of the mass with resection anastomosis was done and 1 patient with an additional sigmoid volvulus which was untwisted with complete debulking of the mass in the other patient. Splenectomy with partial gastrectomy was done in the third patient and resection anastomosis of small bowel with complete excision of the mass was done in the fourth patient.

Two patients were operated for acute appendicitis and they were found to have caecal mass with intestinal perforation. Right hemicolecotomy and ileotransverse anastomosis were performed for both of them. In three
patients, a diagnostic laparoscopy was performed. Harvesting of mesenteric lymph node and liver biopsy were done for all of them.

Of the 15 patients who underwent abdominal exploration, the primary tumour sites were terminal ileum (n=6), caecum (n=2), ileum (n=2), colon (n=2) and lymph nodes /spleen (n=3).

Based on the pathological data; 26 patients were diagnosed as Non-Hodgkin’s Lymphoma (NHL) while 7 patients were diagnosed as Hodgkin’s Lymphoma. The detailed classification was shown in (Table 4).

<table>
<thead>
<tr>
<th>Pathological / Histological diagnosis</th>
<th>Number of patients (n=33)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Non-Hodgkin’s lymphoma (NHL)</td>
<td>26 (78.8%)</td>
</tr>
<tr>
<td>Small lymphocytic</td>
<td>3</td>
</tr>
<tr>
<td>Burkitt lymphoma</td>
<td>7</td>
</tr>
<tr>
<td>Large B-cell</td>
<td>13</td>
</tr>
<tr>
<td>MALT lymphoma</td>
<td>3</td>
</tr>
<tr>
<td>Hodgkin’s lymphoma (HL)</td>
<td>7 (21.2%)</td>
</tr>
<tr>
<td>Nodular sclerosis (Lymph Node biopsy &amp; CT)</td>
<td>5</td>
</tr>
<tr>
<td>Lymphocyte predominant (staging laparotomy)</td>
<td>1</td>
</tr>
<tr>
<td>Lymphocyte depletion (staging laparotomy)</td>
<td>1</td>
</tr>
</tbody>
</table>

Table 4: Pathological and histological results in the studied patients

In the 25 patients who underwent surgical intervention; 19 did not show any post-operative complications. Two patients had wound infection. Four patients were expired; 1 with renal failure, 2 as a result of wound infection and tumor lysis syndrome and 1 because of disseminated metastasis.

Discussion

Despite the existing controversy in diagnosis and treatment of childhood and adolescent lymphoma whether Hodgkin’s or non-Hodgkin’s the role of surgery in the management of these diseases is currently evolving, with the trend towards minimal involvement.

For Hodgkin’s disease staging, laparotomy remains undisputedly the most accurate method to delineate extent of disease precisely. Yet, for therapy of all Hodgkin’s disease patients, young and old, chemotherapy has taken a predominant role. Oncologists thus believe that the precise staging afforded by laparotomy has come to have academic value and no clinical importance.

In our current study, surgical biopsy staging was performed in seven patients with Hodgkin’s disease in with no recorded complications in 5 patients and wound infection in 2 patients. Similar reports were recorded in the literature regarding the superiority of pathologic staging. It was reported that most patients with an equivocal staging should undergo staging laparotomy.

This is contradicted by other data in the literature supported by the concept that the main purpose of staging laparotomy is to identify patients who should be treated with chemotherapy, and because current protocols are mostly chemotherapy-based, laparotomy is unnecessary. Staging laparotomy is used less frequently now. However, the debate regarding the benefits of staging laparotomy still exist as many complications are related to the procedure, although surgical mortality is rare.

In the last few years, chemotherapy has evolved to be the primary modality of treatment for all types of Non-Hodgkin’s lymphomas. Role of radiation therapy and surgery have been regulated to that of symptomatic relief. The dramatic improvement in survival of childhood Non-Hodgkin’s lymphomas can be attributed to multi-agent chemotherapy and improved supportive care. Event free survival of these lymphomas is exceedingly good and thus the current emphasis is to identify newer treatment modalities.

In our current study, patients with non-Hodgkin’s lymphoma huge pelvi-abdominal mass was recorded in 11 (33.3%) patients. Moreover, other abdominal complications due to lymphoma with the need of an emergent surgical intervention were recorded as irreducible intussusceptions in 5 patients, appendicitis in 2 patients and intestinal obstruction in 4 patients with a total percentage of 15.6%, 6.1% and 12.1% respectively.

In a study of 62 patients with abdominal lymphomas treated at Texas Children’s Hospital over a 40-year period, 26 underwent a complete resection. Only 2 complications (small-bowel...
obstruction, wound infection) occurred although a primary anastomosis was performed in all. Of a total of 138 published cases involving complete resection, only 6 complications (5%) were reported. Patients with extensive abdominal disease should undergo a biopsy only followed by the early initiation of chemotherapy.

This data can be compared to our study results that showed an elective abdominal exploration in a total of 4 out of 26 patients with non-Hodgkin’s lymphoma with incisional biopsy for pelvi-abdominal mass in 1 patient. Splenectomy + para-aortic lymph node biopsy & liver biopsy in 3 patients, and 1 patient of total resection of pelvi-abdominal mass+ small bowel resection anastomosis.

The data of our current study showed 2 patients with tumour lysis syndrome that results in hyperuremic nephropathy and renal shut down. This syndrome was also reported in the literature.

We recommend that in patients presenting with extensive intra-abdominal tumour, radical excision is to be contraindicated as the mesenteric root and retro-peritoneum are heavily involved. Attempts at complete excision are associated with a higher complication rate and may lead to a delay in initiation of chemotherapy.

It is also recommended that laparotomy should be limited to cases presented with acute abdomen and limited resectable disease. In the condition of disseminated disease, other alternatives should be applied including bone marrow aspiration, cytological investigation of ascitis/pleural effusion or ultrasound-guided-true-cut needle biopsy of the abdominal mass in order to have less morbidity than laparotomy and allow early initiation of chemotherapy. Despite the benefits of staging laparotomy, many complications are related to the procedure, although surgical mortality is rare.

References


