Myelofibrosis presenting as chronic cholecystitis

Desmin (Dako; 1:100), and S100 protein (Dako; 1:500) was negative. Leucocyte common antigen (Dako; 1:50) stained a few lymphocytes within the abnormal myeloid tissue.

Discussion

The patient had upper abdominal pain characteristic of chronic cholecystitis which responded to cholecystectomy. This implies the pain was originating from the gall bladder; no other source of pain was identified. Myeloid metaplasia or the small gallstone, or both, may have caused the pain. Myeloid metaplasia may have been the sole cause of pain as many gallstones are asymptomatic and there were no histological features of chronic cholecystitis. The gall bladder is rarely biopsied in patients with myelofibrosis and asymptomatic myeloid metaplasia could occur commonly. Thickening of the gall bladder wall can be seen in acute and chronic leukaemia.

The histological diagnosis was relatively straightforward because of the history of myelofibrosis. Without this knowledge, the presence of a pleomorphic infiltrate containing atypical giant cells may have produced an incorrect diagnosis of malignancy. One distinctive feature was that only fibrous tissue was infiltrated and the epithelium and smooth muscle were spared. Also, there was little mitotic activity. Immunohistochemical stains confirmed the correct diagnosis and excluded other possibilities.

As far as we know, there is only one other reported case of myeloid metaplasia of the gall bladder. This occurred in a 71 year old woman with myelofibrosis for 14 weeks, who presented with clinical features of acute cholecystitis. Cholecystectomy was performed and two weeks later she developed acute leukaemia and a pleural effusion containing myeloid cells. She died six weeks after surgery. Unlike this case, "cholecystitis" followed the diagnosis of myelofibrosis, gallstones were not present and there was an acute presentation. In both cases symptomatic involvement of the gall bladder was quickly followed by leukemic transformation, serous effusions and death. There were similar pathological changes in both gall bladders.

In summary, this is the first reported case of myelofibrosis presenting with features of chronic cholecystitis.

We thank Mr S C Toms, Leeds General Infirmary, for photography and Dr C G L Raper, Kingston General Hospital, Hull, for his haematological advice.


Review of clinical activity by microbiologists

A Balfour

Abstract

A data form was devised and used to collect information on clinical cases involving a microbiologist. From the results a relational database management system was created. Of a total of 280 interventions, 137 (49%) were proactive, and in 118 (86%) of these cases the advice given was accepted. The majority of the patients in these cases showed subsequent improvement. Of all the interventions, the given advice was acted upon in 235 (84%), in 22 (8%) it was not and for the remainder this information was not available. This study was a simple method of gaining information on the clinical involvement of the microbiology department of a large city hospital. It provides a reference point from which further research and audit can be based.


Keywords: microbiologist, clinical activity, database.

It is difficult to separate the various components of a medical microbiologist’s activities. One “diary exercise” of a single-handed microbiologist in a district general hospital suggested that approximately one third of the time was spent on each of infection control, clinical activity and laboratory management. Activity in infection control is known to be cost effective but it is harder to show benefit of outcome in clinical activity. This study was designed to determine to what extent microbiologists are involved in the management of clinical cases, and additionally whether their actions actually influence patient outcome.

Methods

A data form (fig 1) was devised and information gathered over a 10 week period. The microbiology department provides a service to its own hospital of approximately 1000 beds, several smaller outlying hospitals, and local general practitioners (GPs). The GP workload is
CONSULTANT CODE Q ON CALL Y/N DATE OF INTERVENTION__

PATIENT DETAILS: SURNAME ___________________________ FORENAME ___________________________
D.O.B. ___________________ PATIENT LOCATION ___________________________

ENQUIER Q INTERACTION BY Q

ADVICE GIVEN WITH RESPECT TO: INFECTION CONTROL Y/N INVESTIGATION Y/N TREATMENT Q
OTHER ___________________________
TIME (MINS) Q ACTED ON Y/N OUTCOME Q

CODE LISTS

<table>
<thead>
<tr>
<th>Enquirer Code List</th>
<th>Treatment Code List</th>
<th>Outcome Code List</th>
<th>Interaction by Code List</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>No advice given</td>
<td>Deterioration</td>
<td>Telephone</td>
</tr>
<tr>
<td>1</td>
<td>Nursing staff</td>
<td>Start</td>
<td>Visit ward/Dept</td>
</tr>
<tr>
<td>2</td>
<td>Medical staff</td>
<td>Change</td>
<td>Letter</td>
</tr>
<tr>
<td>3</td>
<td>Medical staff out</td>
<td>Change</td>
<td>Advice drug reaction</td>
</tr>
<tr>
<td>4</td>
<td>Non Medical staff</td>
<td>Do nothing</td>
<td></td>
</tr>
</tbody>
</table>

Figure 1 Form used for collecting data on interventions involving a microbiologist.

approximately 25% of the total. Four microbiologists were involved in the study: two consultants, one principal bacteriologist, and one senior registrar. Each of them recorded information regarding every clinical intervention with which they were involved. Details recorded were: (1) date of intervention and whether during normal laboratory hours (0900 to 1700 hours Monday to Friday, 0900 to 1200 hours Saturday); (2) patient identity; (3) whether intervention was requested (for example, clinical colleague telephoning for advice) or not (for example, microbiologist telephoning proactively with result of a positive blood culture isolate); (4) whether the intervention was requested, and by whom; (5) mode of interaction (telephone, letter, ward visit); (6) types of advice given (infection control, investigations, treatment, other); (7) if advice was given regarding antimicrobial treatment, whether to stop, start, change, or do nothing; (8) how much time spent in communication of information (excluding time spent in visiting a ward or department); (9) whether advice was taken or not; and (10) if advice was taken, how it influenced patient outcome.

Answers to the latter two questions were derived from patient's notes and from clinicians dealing directly with the patients. While it is accepted that outcome may be a subjective measure, it is believed that the information collected related specifically to the intervention and the patient at that time. The information collected was transcribed from the data forms to a computer relational database management system (Microsoft Access), which was used to collate the results.

Results

A total of 280 interventions were recorded, of which 242 (86%) were during normal laboratory working hours; 38 (14%) were outside normal laboratory working hours. Of the latter, 11 (29%) involved more than four minutes of communication when outside normal laboratory working hours; during normal laboratory working hours; only 15% of interventions involved this length of time (p<0.05 χ² test).

The telephone was the mode of interaction in 82% cases, the remainder being made by visiting a ward or department. Of the interventions, 51% were requested, 76% of these by hospital doctors, 14% by general practitioners, 8% by nursing staff, and 2% by others. The remaining 49% of interventions were proactive.

There were four categories of advice given, with any combination of these possible. Advice was given on treatment in 86% cases, investigations in 27%, infection control in 15%, and for 7% other advice was given such as "refer to Regional Virus Laboratory" or "contact Respiratory Physician".

Where advice was given on antimicrobial treatment, in 40% of cases it was to do nothing, in 28% it was to change antimicrobial treatment, in 22% to start antimicrobial treatment, and in 10% to stop. For these interventions where antimicrobial treatment was advised, 50% of cases were associated with patient improvement, 32% with no change, 8% with a deterioration, and for the remainder, patient outcome was not available.

In 118 (86%) of the 137 proactive interventions the advice given was accepted, and of these 60% were associated with patient improvement, 32% with no change, 4% with a deterioration, and for 4% the outcome was not known. This contrasts with the 13 cases where proactive advice was not acted on. Of these, six (46%) were associated with a deterioration, five (38%) with no change, one (8%) with an improvement, and in the other case the outcome was not known. In the remaining 5% of proactive interventions it could not be ascertained whether advice was acted on or not.

For requested interventions, as for proactive cases, most advice was acted on (117 of 143 cases, 82%). Of these, 56 (48%) were associated with patient improvement. No change was found in 49 (42%), eight (7%) were associated with a deterioration, and for four (3%) the outcome was not known. If requested advice was not acted on (6%), 22% of cases were associated with an improvement, 22% with a deterioration, 44% with no change, and the outcome in the remainder was not known. In 12% of requested interventions, it was not known whether advice given was acted on or not.

Table 1 Patient outcome depending on whether advice was acted on or not

<table>
<thead>
<tr>
<th>Patient outcome</th>
<th>Advice acted on</th>
<th>Advice not acted on</th>
</tr>
</thead>
<tbody>
<tr>
<td>Improvement</td>
<td>126</td>
<td>3</td>
</tr>
<tr>
<td>Deterioration</td>
<td>13</td>
<td>8</td>
</tr>
<tr>
<td>No change</td>
<td>87</td>
<td>9</td>
</tr>
<tr>
<td>Adverse reaction</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Not known</td>
<td>8</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>235</td>
<td>22</td>
</tr>
</tbody>
</table>

A comparison by χ² test of patient improvement or not depending on whether advice was acted on (or not) showed a significant difference at p<0.001.

Balfour
Granulomatous disease in common variable immunodeficiency: effect on immunoglobulin replacement therapy and response to steroids and splenectomy

G P Spickett, J G Zhang, T Green, J Shrimankar

Abstract
A 40 year old white woman with common variable immunodeficiency of four years duration presented with rapidly increasing splenomegaly. Despite high dose, weekly intravenous immunoglobulin, it was impossible to raise the trough serum IgG concentration to within the normal range. While waiting for a diagnostic splenectomy, low dose corticosteroids were started, leading to a decrease in the size of the spleen and an increase in the trough IgG concentration. Both spleen and liver showed non-casing granulomas. Following splenectomy, the corticosteroids were tailed off and the trough IgG was maintained well into the normal range on a reduced, fortnightly dose of intravenous immunoglobulin and a low dose of oral corticosteroid.

Keywords: common variable immunodeficiency, intravenous immunoglobulin, granuloma.

Common variable immunodeficiency (CVID) is a primary immunodeficiency which may occur at any age and is manifest predominantly by antibody deficiency, leading to recurrent bacterial infections. The cause is unknown, but there are widespread immunological abnormalities. About 10% of patients lack B lymphocytes, and some of these patients have now been shown to carry the same molecular defect as has been described in patients with X-linked agammaglobulinemia (XLA); 30% of patients with CVID may have splenomegaly. These patients tend to have a more severe disease clinically and more severe in vitro immunological abnormalities.

A granulomatous disease resembling sarcoidosis has been described in CVID. This condition is not found in XLA, indicating that it is not just a feature of severe or persistent infection, as the infection profiles for CVID and XLA are very similar. The disease behaves clinically very like sarcoidosis and may involve the lung, gut and nervous system. Both Kveim negative and Kveim positive variants have been described. The optimum treatment is unknown, but it is usually responsive to corticosteroids. It is rare for patients with CVID to undergo splenectomy as there is a reluctance to impose an additional immune deficiency on patients with a major pre-existing defect. However, splenectomy may be justified in cases where there is significant hypersplenism or concern about underlying lymphoma, the incidence of which is increased in patients with CVID.