Lymphocytic interstitial pneumonitis associated with autoimmune hepatitis

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A 49-year-old woman was diagnosed as autoimmune hepatitis and started on steroids and azathioprine. Subsequently, she developed fever; chest radiograph showed lower lobe nodular opacities. Bronchoalveolar lavage and transbronchial lung biopsy confirmed the diagnosis of lymphocytic interstitial pneumonitis. [Indian J Gastroenterol 2001; 20: 76-77]

Key words: Chronic hepatitis

Lymphocytic interstitial pneumonitis (LIP) is a reactive lymphoid infiltrate with diffuse or multifocal pulmonary interstitial involvement. It occurs in the setting of several systemic illnesses such as Sjögren's syndrome, myasthenia gravis, chronic hepatitis and primary biliary cirrhosis. LIP has also been described in association with AIDS in children as well as in adults. Since it commonly presents with cough, dyspnea and fever, among patients receiving immuno-suppression it may be misdiagnosed as respiratory tract infection.

A 49-year-old lady presented with fatigue, nausea and jaundice. Total bilirubin was 6.2 mg/dL and transaminases were elevated (AST/ALT 1660/1680 IU/L; normal 0-40); HBSAg and markers for hepatitis A and C viruses were negative. At ultrasonography, liver was of normal size, had altered echotexture, and no mass was seen; there was no biliary dilation. Bilirubin normalized over the next six weeks and AST/ALT were 70 IU/L each; this was accompanied by clinical improvement.

Five months later, she developed nausea, fatigue and jaundice. Total bilirubin was 2.5 mg/dL; transaminases were high (AST/ALT 1248/1344 IU/L) and alkaline phosphatase was 305 IU/L (normal 40-160); prothrombin time was normal. Serology for hepatitis viruses A, B and E, anti-nuclear antibody, antiliver kidney microsomal antibody and antimitochondrial antibodies were negative. Alpha-1-antitrypsin level was normal. Abdominal sonography revealed no hepatomegaly, mass or ascites; hepatic and portal veins were normal. Bilirubin level settled over a few weeks but transaminases remained mildly elevated.

Ten months later, bilirubin and alkaline phosphate were normal and transaminases mildly elevated (AST/ALT 55/76 IU/ L). Liver biopsy revealed chronic hepatitis with piecemeal necrosis and marked activity. There was extensive fibrosis suggestive of cirrhosis. Quantitative iron studies on liver tissue did not reveal iron overload. With a diagnosis of autoimmune hepatitis, the patient was started on corticosteroids and azathioprine. Corticosteroids were tapered and stopped after a month as her diabetes was difficult to control; azathioprine was continued. Transaminases normalized after starting treatment.

One month later, she was admitted with high fever. Leukocyte count was 15,000/µL, bilirubin 1.4 mg/dL, AST/ALT 29/19 IU/L and alkaline phosphatase 64 IU/L. Chest radiograph showed nodular opacities in both lower lobes. There were no respiratory complaints except for occasional breathlessness; chest examination was normal. High-resolution computed tomogram of the chest showed patchy ground-glass densities interpersed with septal thickness. Diffuse nodular opacities were seen in both lungs. Fiberoptic bronchoscopy revealed a normal tracheobronchial tree. Bronchoalveolar lavage showed excess of lymphocytes, but no acid-fast bacilli, fungi, Pneumocystis carinii or other organisms. Transbronchial biopsy revealed moderately dense but patchy infiltrate of lymphocytes in septae, consistent with lymphocytic interstitial pneumonitis (Fig). Pulmonary function test revealed a restrictive defect with FVC of 1.4 L (55% of predicted) with normal FEV/FVC. Azathioprine was continued and 40 mg/day of prednisolone was started. One month later, FVC had improved to 2 L (88% of predicted) and repeat chest radiograph was normal.

The patient was treated with tapering doses of corticosteroids and azathioprine and at follow up after 3 months, liver and lung functions and CT scan of the chest were normal.

LIP is characterized histologically by diffuse infiltration with predominantly small lymphocytes and some plasma cells and histiocytes in the alveolar septa and along the lymphatic vessels. Lymphocytic bronchiolitis with limited involvement of the interstitium is a recognized complication of graft-versus-host disease following bone marrow transplantation. Abnormalities of serum globulin levels are found in more than 60% of patients with LIP.

LIP tends to progress to diffuse fibrosis, though evolution to malignant lymphoma and to lymphomatoid granulomatosis has also been described. The condition
usually responds to high-dose corticosteroids; if these are not successful, cyclophosphamide, chlorambucil or azathioprine may be tried.

Our patient had LIP with autoimmune hepatitis. Patients with autoimmune hepatitis are on immunosuppressive drugs and hence have a high chance of respiratory infection. Since LIP also presents in a similar way, it is often misdiagnosed as an infection.

References

**BOOK REVIEW**

**Operative Procedures in Surgical Gastroenterology.**

This is the latest entrant in the list of books being published by Indian authors with the help of multiple contributors. It underscores the revolution that is taking place in knowledge vis-à-vis the Indian medical literature. The contributors chosen are all distinguished surgeons in the field of Surgical Gastroenterology. Professor Kaushik himself has been at the forefront of the development of this specialty in India.

The book has many strengths. It is written in “terse lucid prose” (as aptly described in the Foreword by Prof. Michael Hobsley), which is quite effective, with operative steps being elucidated in an easy-to-follow point-wise style. Contents are well organized, with frequent highlighted headings, which is helpful in navigating the book. Surprisingly, unlike most books with contributions from many authors, this one is not uneven, but all the authors have followed the same pattern. For this, the credit should go to the editor.

The book is organized into several chapters dealing with different surgical techniques. Most of the chapters on esophageal, gastric, hepatobiliary, pancreatic and colorectal surgery are well written. Some of the thoughtful chapters, like the ones on partial cholecystectomy, segment III cholangiojunoscopy, cholecystectomy in Mirizzi’s syndrome, Roux-en-Y loop reconstruction, and gastrojejunal fistula will prove especially useful to young surgeons, as information on these topics is rather sparse in literature like this. There is also a comprehensive index at the end. Overall, it is an easy read.

The book is not without its share of flaws and weaknesses, some of which are quite obvious. Diagrams are few and far in-between; more diagrams would have gone a long way in enlightening the reader about the finer nuances of the operative steps. Some of the authors have been rather reticent in sharing detailed analysis of their own data. For example, the chapter on Transhiatal Oesophagectomy, by Prof. T K Chattopadhyaya and a colleague, whose vast experience in this procedure is perhaps second to none in the Indian subcontinent, has data limited to operations done before 1996. Few will agree with the list of benign conditions for radical gastrectomy (page 41). Such shortcomings convey an impression of a missing stamp of authority from senior authors. Budding and aspiring surgeons would have appreciated the use of highlighting, shaded boxes directing the reader’s focus, and “yellow flagging” of the areas and pitfalls in these advanced procedures and tips on how to avoid them. References are not marked in the text, leading to a rather difficult search at the end of the chapters. Some references are incomplete (on pages 16, 38, 40, 63 and 163). There is very little here that is new or stimulating; for a connoisseur looking for cordon bleu delights, the book comes across like fast food: neat and adequate but slightly bland.

Notwithstanding these reservations, the book may quickly become dog-eared and worn from frequent use. The book is, as mentioned in the Preface, meant to be "a support": a type of companion or aid to memorizing for the "young faculty, an MCh / MS postgraduate and a practitioner of surgery to remember basic and essential steps." No doubt this purpose is served, but readers will have to read between the lines to understand why a procedure is performed in a particular way.

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