Clinical Aspects of Liver Diseases

38 Systemic diseases and the liver

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Systemic diseases originating in the *haemopoietic* or *lymphatic* systems and those belonging to the category of *rheumatic diseases* can affect the liver directly or indirectly.

Haematopoietic system
1. ***Direct effects on the liver***
   = pathological extramedullary haemopoiesis in the liver
   = disturbed liver haemodynamics following haemolysis
2. ***Indirect effects on the liver***
   = reduced defence against infections facilitates bacterial, viral or mycotic liver damage
   = toxic liver damage caused by drugs (e. g. cytostatics, immunosuppressants)
   = graft-versus-host reaction in bone-marrow transplants

Lymphatic system
1. ***Direct effects on the liver***
   = pathological formation and deposition of lymphocytes or lymphoblasts with formation of infiltrates or focal lesions
   = cholestasis following mechanically mediated biliary dyskinesia
2. ***Indirect effects on the liver***
   = reduced defence against infections facilitates bacterial, viral or mycotic liver damage
   = toxic liver damage caused by drugs (e. g. cytostatics, immunosuppressants)

Rheumatic diseases
1. ***Direct effects on the liver***
   = rheumatism-related inflammatory or immunologically induced intrahepatic vasculitis with sequelae
   = non-specific reactive hepatitis
2. ***Indirect effects on the liver***
   = toxic liver damage caused by drugs (e. g. antirheumatic agents, immunosuppressants)

Tab. 38.1: Relationship of the liver to the haematopoietic and lymphatic systems and to the category of rheumatic diseases

1 Systemic haematological diseases

1.1 Extramedullary haemopoiesis

► Intrahepatic haemopoiesis is a **physiological process** in foetuses and neonates. From 6th to 24th week of pregnancy, haemopoiesis takes place in the liver (and spleen) in a diffuse manner within the sinusoids. Thereafter, focal haemopoiesis may still continue in the liver up to about the second week of life.

Myeloid metaplasia beyond this physiological endpoint of intrahepatic haemopoiesis is regarded as a **pathological event.** Haemopoietic foci are seen in the sinusoids, in Disse's spaces and sometimes to a minor degree in the portal fields; they consist of erythropoietic and myeloproliferative precursors as well as polynuclear giant cells of the megakaryocyte type. This variety of cells provides important evidence in histological differential diagnosis for excluding leukaemic infiltrates and mononuclear hepatitis. Myeloid metaplasia accompanies displacement of the bone marrow, e. g. in osteomyelofibrosis, bone-marrow carcinomatosis and myeloproliferative diseases. Occasionally, megakaryocytes are also present in the liver. Naphtol-AS-D chloracetate esterase-positive cells of granulopoiesis are a striking feature in this context. Hepatomegaly is generally found. Ascites can develop due to portal hypertension. (s. fig. 38.1)

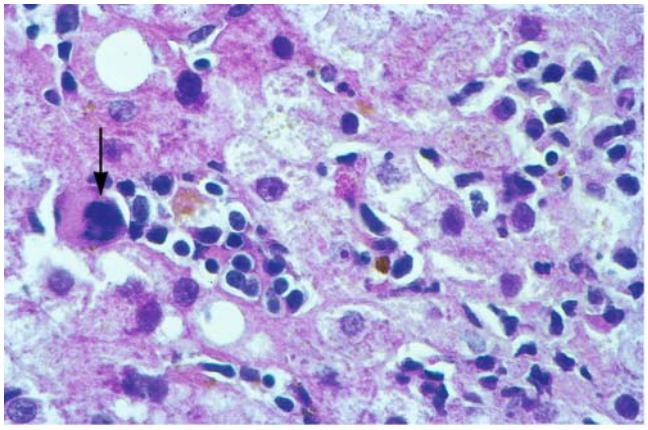

Fig. 38.1: Extramedullary haemopoiesis in the liver with erythropoiesis precursors and intrasinusoidal megakaryocyte (↑) due to the so-called marrow-replacement syndrome or chronic myeloproliferative disease (HE)

1.2 Acute leukaemia

In acute myeloid leukaemia or lymphatic leukaemia as well as in acute leukaemic episodes in non-Hodgkin lymphoma, involvement of the liver may only be detectable clinically by the presence of hepatomegaly and subicterus. • **Laboratory parameters** usually show slightly elevated transaminase as well as bilirubin values, and distinct cholestasis is occasionally observed. (7) Acute hepatic failure can occur during the course of acute leukaemia. (1, 8, 26, 65) • **Histologically,** there are massive, yet uniform blast-cell infiltrates; these are found mainly within the portal fields in acute lymphatic leukaemia (about 95%) and within the sinusoids in acute myeloid leukaemia (about 75%). • Involvement of the liver is of no consequence with regard to the underlying disease and its **therapy**. Secondary infections require systemic treatment with antibiotics and/or antimycotics.

1.3 Myeloproliferative syndrome

The term myeloproliferative syndrome encompasses the following: (*1.*) chronic myeloid leukaemia, (*2.*) idiopathic osteomyelofibrosis and sclerosis, (*3.*) polycythaemia vera, and (*4.*) essential thrombocytosis. Inter-