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Pathology of peliosis

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Abstract

Peliosis is a pathological entity characterized by the gross appearance of multiple cyst-like, blood-filled cavities within parenchymatous organs. Peliosis has been related to several underlying debilitating illnesses such as tuberculosis, hematological malignancies, the acquired immunodeficiency syndrome (AIDS), and post-transplant immunodeficiency, as well as intravenous drug abuse, chronic alcoholism, and in conjunction with the intake of oral contraceptives or steroids. The classical pathoanatomical concept is based upon the opinion that peliosis exclusively develops in organs belonging to the mononuclear phagocytic system (liver, spleen, bone marrow, and lymph nodes). However, a paucity of studies indicates that other organs such as lungs, parathyroid glands, and kidneys may be affected too. Concerning the underlying pathogenetic mechanisms of onset and maintenance of peliosis, the morphological data obtained by different investigators suggest that there is more than one path of formal pathogenesis (e.g., congenital malformation of vessels manifesting under altered local intravascular pressure conditions, acquired vascular disorder triggered by toxic noxae, active proliferation of vessels corresponding to the benign end on the spectrum of neoplastic vascular lesions). In the liver, at gross inspection, the peliotic lesions give the cut sections a ''swiss cheese'' appearance. Microscopically, two different types of peliosis can be distinguished in the liver: (1) ''parenchymal peliosis'' consisting of irregular cavities that are neither lined by sinusoidal cells nor by fibrous tissue, and (2) ''phlebectatic peliosis'' characterized by regular, spherical cavities lined by endothelium and/or fibrosis. One of the differential diagnoses that most closely resembles peliosis hepatis is secondary hepatic congestion due to veno-occlusive disease or the Budd–Chiari syndrome. In the spleen, the peliotic lesions may be arranged sporadically, disseminated, or in clusters in an uneven distribution pattern. Histologically, the cavities show frequently well-demarcated margins that may appear focally lined by sinusoidal endothelium, or totally lack a clear cell lining. Differential diagnoses are hemangiomas and involvement of the spleen in hairy-cell leukaemia. Since the disease may culminate in spontaneous rupture of the affected organ and thus may mimick a violent death at autopsy, peliosis is far more than just another morphological curiosity. Awareness of peliosis at autopsy as well as an appreciation for the histopathological changes in less characteristic or advanced cases may become an important issue for both the forensic and clinical pathologist.

Keywords: Peliosis; Liver; Spleen; Mononuclear phagocytic system; Intraperitoneal hemorrhage; Sudden death; Autopsy; Forensic histopathology

1. Introduction

Peliosis is a comparatively rare pathological entity characterized by the presence of multiple blood-filled cavities within parenchymatous organs. The word ''pelios'' is greek, meaning *blackish–bluish with sugillation*, and was first used by Wagner in 1861 to describe the gross appearance of the lesions on cut surfaces of the liver [1]. The diagnosis of peliosis may be occasionally established at autopsy by the unique gross appearance of multiple cyst-like, blood-filled cavities on cut-surfaces of the affected organ, but usually the disease is a histological diagnosis [2–7]. Peliosis has been

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related to several underlying debilitating illnesses or medications, and different theories have been proposed concerning the pathogenesis of this unique disorder over the years. While some investigators favour congenital malformation of vessels or microcirculatory disturbances manifesting under altered local intravascular pressure conditions as responsible for the development of the disease [4,8–10], others believe that an acquired vascular disorder triggered by toxic noxae may play a key role in the pathogenesis [11,12]. However, none of the chronic or toxic conditions previously described in association with peliosis or the pathogenetic mechanisms they may induce can satisfactorily explain the pathological findings in all cases and organs involved, respectively, and the morphological data obtained by different investigators suggest that there is more than one path of formal pathogenesis to the onset and maintenance of peliosis.

The classical pathoanatomical concept is based upon the opinion that peliosis exclusively develops in organs belonging to the mononuclear phagocytic system (MPS, formerly known as the ''reticuloendothelial system'') and therefore only affects the liver, spleen, bone marrow, and lymph nodes [12,13]. However, a paucity of studies indicates that other organs such as lungs, gastrointestinal tract, parathyroid glands, pituitary, pancreas, adrenals, and kidneys may be affected by the disorder too [14–18].

Apart from the pathology of peliosis of the liver, the vast majority of patho-anatomical textbooks and manuals devote no attention to the distinctive morphological features of the disease. This review summarizes the pathological features of peliosis, pointing out towards the forensic pathological significance of the disease. It was neither the objective of the present work to give an extensive overview on the available literature on peliosis nor to discuss clinical aspects of the disease. For the latter, it is referred to the comprehensive clinical literature related to peliosis.

2. Peliosis of the liver

In 1916, Schoenlank introduced the term peliosis hepatis into the literature describing the case of a 33-year-old woman who died of miliary tuberculosis [19]. In his case report, nearly 90 years ago, Schoenlank described the gross pathology of the liver as follows: ''Underneath Glisson's capsule, in a diffuse pattern, bluish–reddish–blackish spots that have a circular appearance and measure up to 0.15 cm are visible. On cut surfaces, these spots come out into the open as bleedings (. . .). One cannot squeeze out the blood, it is apparently clotted (. . .). All lesions give the impression that they have developed at the same time and are fresh of origin (. . .)''. At microscopic examination, Schoenlank noticed relatively well-defined margins of the lesions that were not lined by any particular lining cells but by a delicate fibrin network. This author theorized some type of inundation of toxic noxae into the portal circulatory system on the one hand or agonal right heart failure with subsequent stasis of blood in the liver sinusoids on the other hand as responsible for the observed liver pathology.

Since Schoenlank's publication, numerous case reports and small series of cases have appeared in the literature, describing the macroscopical and histological aspects of peliosis hepatis. On cut surfaces, multiple, randomly distributed, blood-filled cystic spaces can frequently be seen at gross inspection, giving the section a ''swiss cheese'' appearance (Fig. 1). Microscopically, two different types of lesions have been described [20]. The first type is

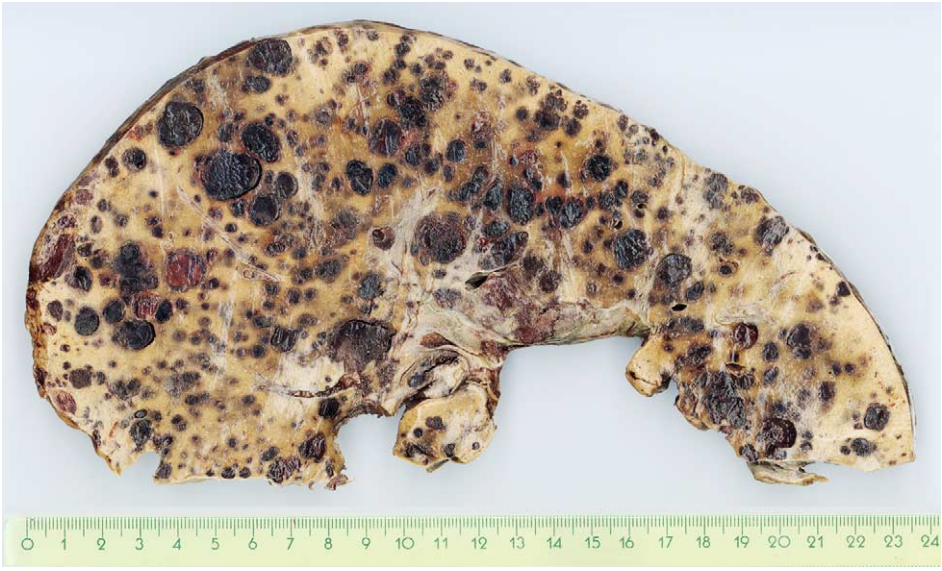

Fig. 1. Peliosis hepatis. Cut surface of liver parenchyma displaying multiple blood-filled cystic spaces that give the section a ''swiss cheese'' appearance.