

Icing Sugar Spleen/Perisplenitis Cartilaginea: A Case Report

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Abstract

Spleen, being the largest organ of the mononuclear phagocytic system is involved in many systemic inflammations, generalized haematopoietic disorders as well as metabolic disorders. Perisplenitis, also known colloquially as “icing sugar spleen”, is a common autopsy finding. The capsule of the spleen becomes nodular, thickened and fibrotic and it appears as though the spleen has been dipped in white icing. Perisplenitis is seen as creamy yellow to white exudates or firm glistening “icing-like” plaques on the capsular surface, indicating active acute or regressed infection, respectively. It is usually secondary to generalised peritonitis or extension from local infection. It has been reported as part of Curtis-Fitz-Hugh syndrome or a common accompaniment to cirrhosis of the liver. Massive splenomegaly with sugar icing spleen is also seen in chronic myeloid leukemia and may be seen in sickle cell anaemia. Here we present a case of perisplenitis in a 65 years old male patient as an incidental autopsy finding.

Keywords: Perisplenitis cartilaginea; Icing sugar; Fibrous perisplenitis; Hyaline perisplenitis

Introduction

Spleen is the largest organ of the mononuclear phagocytic system and is involved in all systemic inflammations, generalized haematopoietic disorders as well as metabolic disturbances [1,2].

Perisplenitis, also known colloquially as “icing sugar spleen”, is a common autopsy finding. The capsule of the spleen becomes nodular, thickened and fibrotic, and it appears as though the spleen has been dipped in white icing. Over time, calcification may supervene. Perisplenitis can complicate any inflammatory process in the abdomen or pelvis and splenic sepsis, and it is also seen in patients with chronic obstructive pulmonary disease. It has been reported as part of Curtis-Fitz-Hugh Syndrome [3].

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Irregular pale tan plaques of collagen over the purple capsule known as “sugar icing” or “hyaline perisplenitis” which follows the splenomegaly and/or multiple episodes of peritonitis that are a common accompaniment to cirrhosis of the liver [5]. Splenic infarcts and perisplenitis may give rise to left hypochondriac pain [6].

Case Report

During an autopsy on a 65 years old male patient, incidentally we came across a mildly enlarged spleen of size 10 × 7.5 × 5 cm. External surface showed multiple, variable sized, whitish nodules and plaques ranging from 2 mm to 1 cm over the convex surface of the splenic capsule and without involvement of splenic parenchyma as seen on cut surface (Figure 1).

On microscopy, these nodules and plaques were composed of closely packed hyalinised collagen fibres over the capsule without involvement of splenic parenchyma (Figure 2).

Discussion

Many surgical pathologists approach the spleen with trepidation, thus indirectly reinforcing the old axiom that the spleen is an organ of

mystery. The reason may be related to the fact that many pathologists initially learn splenic pathology at post-mortem examination; at autopsy, spleen frequently is autolysed, and the normal histologic landmarks are obscured [7]. Perisplenitis is very common incidental observation at post-mortem examination, remain unexplained in pathogenetic terms. It takes the form of pearly white thickening of splenic capsule to form irregular plaques or occasionally, a dense rind on the surface of the organ. The convex surface usually more severely affected than the concave, medial aspect of the spleen [8]. Perisplenitis is an active or regressed phase. When active, part or the entire capsule

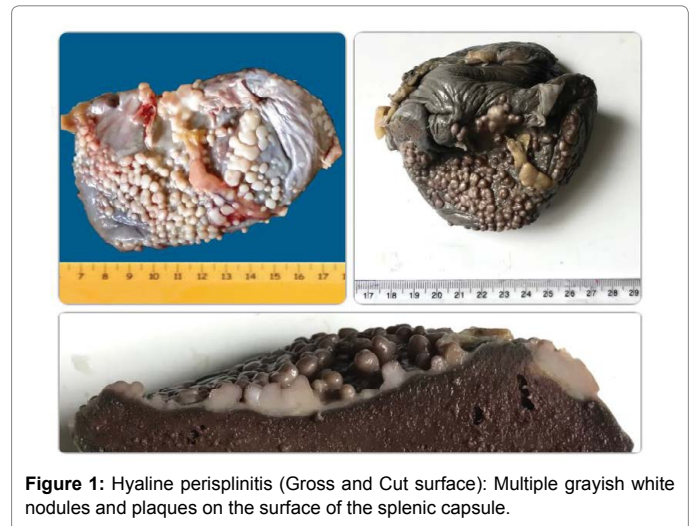


Figure 1: Hyaline perisplenitis (Gross and Cut surface): Multiple grayish white nodules and plaques on the surface of the splenic capsule.

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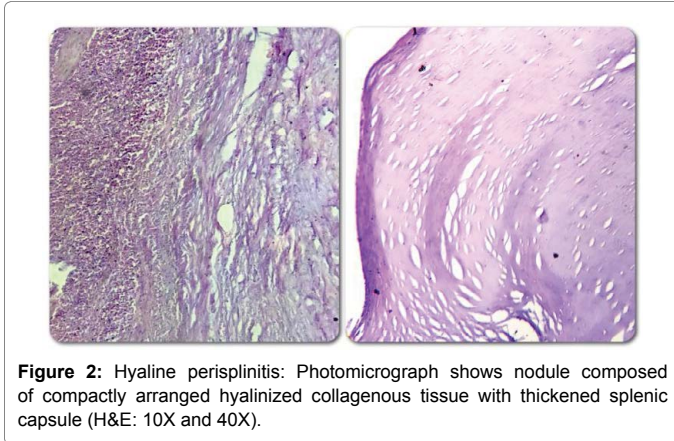


Figure 2: Hyaline perisplenitis: Photomicrograph shows nodule composed of compactly arranged hyalinized collagenous tissue with thickened splenic capsule (H&E: 10X and 40X).

is covered with fibrin which hides the surface sheen and roughens the capsule. The cause is generalised peritonitis or local inflammation spreading from neighbouring organs. Alternatively, it may come from within the spleen. Splenitis, in the sense of infection, is uncommon. Perisplenitis is more likely to be a reaction to thrombosis or infarction. The final stage of perisplenitis is when exudate has long ago been organised, leaving a pure white smooth or wrinkled thickening generally local and often multiple (hyaline perisplenitis). The plaques are several millimetres thick and often have calcium deposits which cannot be cut through. Even greater and quite diffuse thickening is caused by polyserositis; the capsule displays the sugar icing effect seen also around the liver and in the pericardium [9]. Massive splenomegaly with sugar icing spleen is also seen in chronic myeloid leukemia [7]. In the sickle cell anaemia, focal accentuation of capsular fibrosis leads to a "sugar-coated" appearance in the spleen [10]. Perisplenitis cartilaginea is the name derived from hyaline transformation of connective tissue where there is loss of fibrous appearance due to high degree packing of collagen fibers and accumulation of nonfibrillar components (giant proteoglycans e.g. seen in cartilaginous tissue) there by having homogenous translucent character. Capsular fibrosis of the spleen is typically asymptomatic and of little clinical significance. Its importance is in recognizing that it is a benign indolent process and in not confusing it with primary or metastatic malignant disease [11].

Conclusion

It has been reported as part of Curtis-Fitz-Hugh syndrome or a common accompaniment to cirrhosis of the liver. Massive splenomegaly with sugar icing spleen is also seen in chronic myeloid leukemia and may be seen in sickle cell anaemia. Here we present a case of perisplenitis in a 65 years old male patient as an incidental autopsy finding. No MRI was done prior to autopsy. Perisplenitis was an incidental finding during autopsy.

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