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Isolated Peliosis Lienis – A Case Report and Literature Review

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Abstract

Peliosis is a condition characterized by presence of multiple blood filled cysts within the parenchyma of a solid organ. Peliosis affecting spleen is usually seen in association with peliosis of liver.

Isolated splenic peliosis "peliosis lienis" is a unique phenomenon. At Government Medical College, Thiruvananthapuram, Kerala, India we had one such case.

Dead body of a 27 year old male was brought for medico legal autopsy with history of being found dead in the morning. He had a history of mild abdominal pain on the previous day. Autopsy revealed splenic peliosis with rupture of spleen and haemoperitoneum.

Keywords: Splenic peliosis; Peliosis hepatis; Splenic rupture; Haemoperitoneum

Introduction

Peliosis is a rare condition characterized by multiple cysts like blood filled spaces within the parenchyma of a solid organ [1]. It commonly affects liver or liver and spleen together are usually involved [2]. When it affects liver, it is hepatic peliosis or peliosis hepatis. When it affects spleen alone, it is splenic peliosis or peliosis splenis or peliosis lienis [1]. Isolated splenic peliosis is extremely rare.

When peliosis occurs in isolation in spleen, the condition is often asymptomatic [2]. It is a potentially lethal condition and present with splenic rupture and haemoperitoneum. It may be discovered incidentally on imaging for other indications or at autopsy [3].

Case Report

A 27 year old male was working as a hotel boy in a small restaurant in a village at Thiruvananthapuram, Kerala state. One day after his daily work, he came home in the afternoon with complaints of mild and vague abdominal pain.

There were no other associated disturbances or symptoms. After a normal dinner he went to bed as usual. The next day morning, he was found dead in his bed.

After preliminary investigations and inquest procedures by the police, the body was brought for medico legal autopsy to Government Medical College, Thiruvananthapuram.

Autopsy Findings

Body was of a moderately built and under nourished male of height 154 cm and weight 40 Kg on external examination. There were no injuries on the body. During internal examination relevant findings were observed.

Spleen was found enlarged and weighed 210 g. It was soft with multiple areas of sub capsular hemorrhages.

Multiple blood filled cysts of varying sizes were found in the parenchyma of spleen. One of which was found associated with an area of capsular rupture (Figure 1).

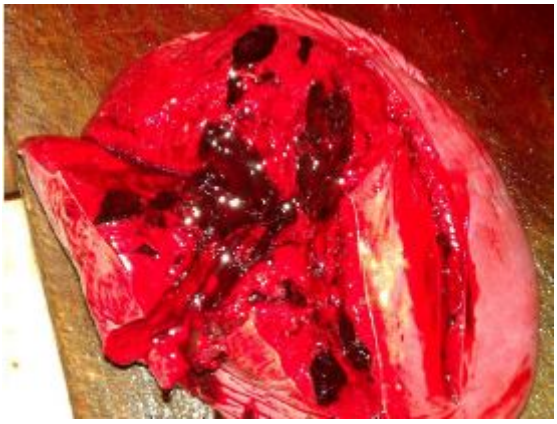

Figure 1: cut section of spleen showing multiple blood filled cavities

No other abnormality was noted in any of the organs. Samples of viscera and blood subjected to chemical analysis revealed no poison.

All other organs including liver were pale, otherwise appeared normal. Heart showed subendocardial haemorrhages.

Peritoneal cavity contained 2000 ml of fluid blood and 200 g of blood clot (Figure 2).

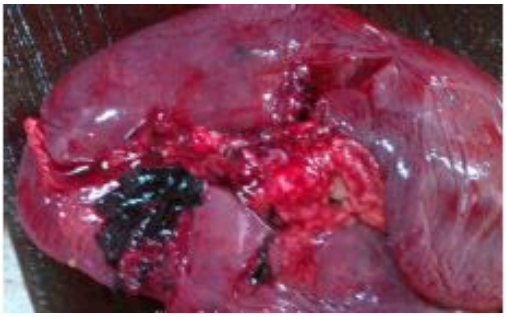

Figure 2: showing capsular rupture of peliotic cavity

Histopathological examination of the spleen showed multiple areas of hemorrhages (Figure 3).

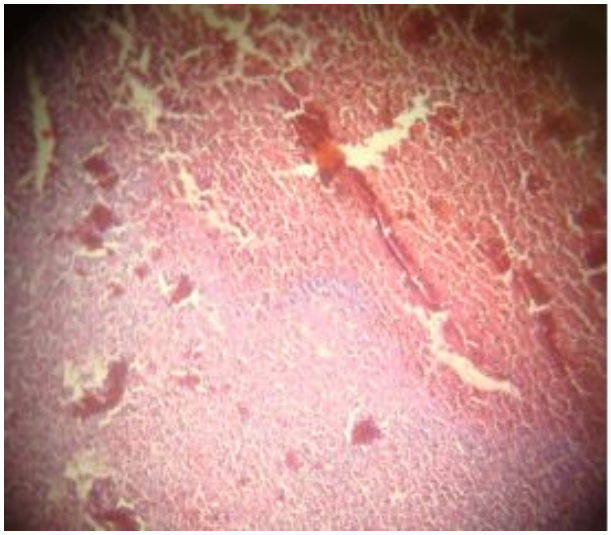

Figure 3: Low power view of spleen showing multiple haemorrhagic areas

Discussion

Peliosis is an unusual benign disease characterized by presence of irregular cystic blood filled cavities in the parenchyma of solid organs [2]. Less than hundred cases have been so far reported in literature. The term 'peliosis' originated from the Greek word 'pelios' meaning dusky or purple. It is the gross macroscopic appearance of the organ gave this name to this condition [4]. The name was first used by Wagner [5]. Peliosis develops in organs belonging to mono nuclear phagocyte system mainly liver and spleen, it is also seen in bone marrow and lymph nodes [6]. Certain other studies revealed that organs such as lungs, parathyroid glands, adrenals and kidneys may also be affected [7]. Peliosis was first described in liver i.e. peliosis hepatis by Schoenland [8]. Isolated splenic peliosis is extremely rare and was first reported in 1978 [9]. Until then peliosis of spleen was thought to occur only in association with peliosis hepatis [10]. Tada et al. examined spleen in 1200 autopsy cases over a period of three years from 1977 to 1980 and peliosis was found in 10 cases [11]. It was confined to spleen only in 8 cases. Its prevalence is seen in less than 1% cases of autopsy [11].

Aetio-pathogenesis of splenic peliosis is not fully understood. It may be seen in association with various conditions and various authors have reported these conditions. Splenic peliosis has been related to debilitating diseases like hematological malignancies, tuberculosis, acquired immune deficiency syndrome (AIDS), mono clonal gammopathies like multiple myeloma, other malignancies like Hodgkin's disease, seminoma and diabetes mellitus in end stage renal failure on peritoneal dialysis and erythropoietin therapy in cirrhosis of liver and post liver transplantation [7,12-18]. Drugs and toxins including chronic alcoholism, prolonged use of drugs like anabolic steroids, oral contraceptive pills, tamoxifen, azathioprim, danazol etc were also seen associated with splenic peliosis. Peliosis was also reported in puerperium after normal pregnancy [1,19-24].

This disease is more common in males [11] with a male female ration of 1.7:1. The age distribution ranges from 14 to 82 years [6].

Pathogenesis is still unclear; several processes in pathogenesis have been postulated by various authors. Tada reported that para follicular areas are the most common sites of the lesion [11]. Damage to sinusoidal walls is considered as the primary event in pathogenesis [1,18]. Local micro circulatory disorders manifested by altered local intra vascular pressure in the spleen may be responsible for Peliosis associated with vascular lesion [20-25]. Another view suggested by Tsakos is that these lesions represent vascular malformations which manifests under locally altered intravascular pressure conditions which may be congenital or acquired [7]. Some other reports described immune complex deposits in the vicinity of peliotic lesion [26,27]. Etzion et al. reported a case of a traumatic rupture of spleen in splenic peliosis with increased macrophage activity indicating immune complex basis of the condition [28]. Gugger et al. detected deposits of Ig G with complement C3 around peliotic areas [27]. This is supportive of the hypothesis suggested by Etzion. There are varying stages for this disease ranging from cystic dilatation to early organization of blood clot within the lesion and finally fibrous scar formation as described by some pathologists [29].

Splenic peliosis is usually asymptomatic or have mild abdominal pain. This condition may be diagnosed as an incidental finding during autopsy in adult [30]. Diagnosis can be made macroscopically. Numerous cysts like blood filled cavities can be demonstrated on cut section. Ultrasound scanning (USS) or contrast enhanced computed tomography scanning (CECT) or Magnetic resonance image (MRI) scanning of abdomen for some other abdominal pathology may reveal this condition in live subjects. It can mimic metastasis on CT abdomen [31].

Haemangiomas and hairy cell leukemia are the conditions which should be differentiated from splenic peliosis [32]. Although splenic peliosis is termed as "benign condition" life threatening complications can occur. The risk of these events has not been quantified. When peliotic cysts rupture either spontaneously or by trivial trauma resulting in haemoperitoneum and prove to be fatal [3]. Celebrezze et al. reported seven documented cases of spontaneous splenic rupture associated with isolated splenic peliosis [15]. There are no published guidelines on managing patients with suspected splenic peliosis, as this condition is so rare [5]. Splenectomy is the treatment of choice in patients with splenic rupture. Haemorrhage associated with splenic peliosis is amenable to surgery if diagnosed and treated promptly [33].

Prophylactic splenectomy can also be advised in cases of incidental diagnosis of splenic peliosis.

Medico Legal Significance

The significance of peliosis lienis arises in many situations. There is a potential of spontaneous splenic rupture in persons with peliosis, which may sometimes come under false allegation of violent death. Blunt trauma leading on to rupture of spleen may sometimes lead at a false conclusion of sudden natural death.

The same can be applied in workplace also. A trivial trauma can cause rupture of peliotic spleen. Trauma could be result of a personal accident or by a criminal act. In both situations trivial blow in the abdominal region may not cause significant external injury. This causes difficulties for the investigating officer to find out the manner of death.

Conclusion

Peliosis lienis is an extremely rare condition and may remain asymptomatic. But it can be life threatening due to the rupture of the organ either spontaneously or following a trivial injury. It is usually associated with various debilitating diseases or with drug intake. In the instant case, no associated disease is revealed in histopathological examination or drug intake from detailed investigation.

When a patient attends the emergency department with acute abdomen with or without hemorrhagic shock splenic peliosis should also be suspected. Prompt diagnosis and emergency splenectomy can be lifesaving in such situation

Key Points

- Isolated splenic peliosis, characterized by presence of multiple blood filled cysts within organ is a unique phenomenon.
- Less than hundred cases have been so far reported.
- Its pathogenesis is unclear and may be seen in association with various conditions.
- It is usually asymptomatic, but life threatening complications like haemoperitoneum occurs it ruptures.

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Reference

1. Lashbrook DJ, James RW, Phillips AJ, Holbrook AG, Agombar AC, et al. (2006) Splenic peliosis with spontaneous Splenic rupture: report of two cases. BMC Surg 6: 9.
2. Davidson J, Tung K (2010) Splenic Peliosis: an unusual entity. Br J Radiol 83: e126-e128.
3. Warfel KA, Ellis GH (1982) Peliosis of the Spleen: report of a case and review of the literature. Arch Pathol Lab Med 106: 99-100.
4. Kleinig P, Davies RP, Maddern G, Kew J (2003) Peliosis hepatitis: central 'fast surge' ultrasound enhancement and multislice CT appearances. Clin Radiol 58: 995-998.
5. Singh-Ranger G, Rajarajan N, Aftab S, Stoker D (2007) Splenic peliosis – a potentially fatal condition which can mimic malignancy. Int Semin Surg Oncol 4.
6. Ribeiro-Junior MAF, Zanco GL, Sala MAS, El-Ibrahim R, Saad WA, et al. (2011) Splenic peliosis – a case report. ABCD Braz Archiv Digest Sur 24: 186-187.
7. Tsokos M, Erbersdobler A (2005) Pathology of Peliosis. Forensic Sci Int 149: 25-33.
8. Gushiken FC (2000) Peliosis hepatis after treatment with 2- chloro-3'-deoxyadenosine. South Med J 93: 625-626.
9. Joseph F, Younis N, Haydon G, Adams DH, Wynne S, et al. (2004) Peliosis of the spleen with massive recurrent haemorrhagic ascites despite splenectomy and associated with elevated levels of vascular endothelial growth factor. Eur J Gastroenterol Hepatol 16: 1401-1406.
10. Lacson A, Berman LD, Neiman RS (1979) Peliosis of the spleen. Am J Clin Pathol 71: 586-590.
11. Tada T, Wakabayashi T, Kishimoto H (1983) Peliosis of the Spleen. Am J Clin Pathol 9: 708-713.
12. Diebold J, Audouin J (1983) Peliosis of the spleen. Report of a case associated with chronic myelomonocytic leukaemia presenting with spontaneous splenic rupture. Am J Surg Pathol 7: 197-204.
13. Gabor S, Back F, Csiffary D (1992) Peliosis lienis: Uncommon Cause of Rupture of the Spleen. Path Res Pract 188: 382-383.
14. Fidelman N, LaBerge JM, Kerlan RK (2002) SCVIR 2002 Film Panel case 4: massive intraperitoneal haemorrhage caused by Peliosis hepatis. J Vasc Interven Radiol 13: 542-545.
15. Celebrezze JP, Cottrell DJ, Williams GB (1998) Spontaneous Splenic rupture due to isolated splenic peliosis. South Med J 91: 763-764.
16. Lam KY, Chan AC, Chan TM (1995) Peliosis of the spleen: possible association with chronic renal failure and erythropoietin therapy. Postgrad Med J 71: 493-496.
17. Bleiweiss IJ, ung SN, Goodman JD (1986) Peliosis of the spleen in a patient with cirrhosis of the liver. Arch PatholLab Med. 110: 669-671.
18. Raghavan R, Alley S, Taw k O, Webb P, Forster J, et al. (1999) Splenic peliosis: a rare complication following liver transplantation. Dig Dis Sci 44: 1128-1131.
19. Garcia RL, Khan MK, Berlin RB (1982) Peliosis of the spleen with rupture. Hum Pathol 13: 177-179.
20. Shimono T, Yamaoka T, Nishimura K, Naya M, Hojo M, et al. (1998) Peliosis of the spleen: splenic rupture with intraperitoneal hemorrhage. Abdom Imaging 23: 201-202.
21. Taxy JB (1978) Peliosis: a morphologic curiosity becomes an iatrogenic problem. Hum Pathol 9: 331-340.
22. Lo CM (2007) A case of splenic peliosis associated with pathologic rupture. J Emer Med 14: 174-178.
23. Penalver JF, Somolinos N, Villanueva C, Sanchez J, Monteagudo D, et al. (1998) Splenic peliosis with spontaneous splenic rupture in a patient with immune thrombocytopenia treated with danazol. Haematologica 83: 666-667.
24. Patricot LM, Dumont M, Duvernois JP, Baulieux J, Mercatello A (1986) A case of hepatic and splenic Peliosis occurring in the

puerperium after normal pregnancy. J Gynecol Obstet Biol Reprod 15: 321-326.

25. Tsokos M, Puschel K (2004) Isolated peliosis of the spleen: report of 2 autopsy cases. Am J Forensic Med Pathol 25: 251-254.
26. Korner M, Gebbers J (2002) Peliosis of the spleen and haemolytic anaemia. Histopathology 41: 179-180.
27. Gugger M, Gebbers J (1998) Peliosis of the spleen: an immune complex disease. Histopathology 33: 387-389.
28. Etzion Y, Beharroch D, Saidel M, Riesenberg K, Gilad J, et al. (2005) Atraumatic rupture of the spleen associated with hemophagocytic syndrome and isolated splenic peliosis. APMIS 113: 555-557.
29. Damjanov I, Linder J (2009) Anderson's pathology (10th edition). Mosby, United States.
30. Weidner N, Cote RJ, Suster S, Weiss LM (2003) Modern surgical pathology (2nd edition). Elsevier, London.
31. Engel P, Tjalve E, Horn T (1993) Peliosis of the spleen associated with a paraganglioma. Case report. Acta Radiol 34: 148-149.
32. Abbott RM, Levy AD, Aguilera NS, Gorospe L, Thompson WM (2004) From the archives of the AFIP: primary vascular neoplasms of the spleen: radiologic-pathologic correlation. Radiographics 24: 1137-1163.
33. Kohr RM, Haendiges M, Taube RR (1993) Peliosis of the Spleen: A Rare Cause of Spontaneous Splenic Rupture with Surgical Implications. Am Surg 59: 197-199.