Case Report

A Rare Abdominal Mass, A Case Presentation

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Abstract

An ectopic spleen is a very rare presentation, the incidence being 20 – 30 %. In our context, we have few reported cases. Most of them present as asymptomatic and can be managed conservativrely.But when presented with massive splenomegaly with a threat to rupture, torsion and discomfort splenomegaly is indicated.

So far, till date only one case has been reported of operated splenectomy, this being the 2nd one In Nepal.

The ectopic spleen is common in female. The Male: Female ratio being 7:1 and is more in between 20 - 40 years of age & rarely encountered in children. It is due to the lax attachments of the spleen to the retroperitoneum with long splenic vessels such that spleen wanders in the abdomen hence ectopic spleen is also called wandering spleen.

The failure of the dorsal mesogastrium to fuse with posterior abdominal wall during embryonic period results in the failure of the formation of the suspensory ligament. The result is unusually a long splenic pedicle making spleen to wander. It has been postulated that acquired defect in splenic attachment may occur in multiparous women secondary to hormonal changes during pregnancy and associated abdominal laxity.

The diagnosis is difficult but duplex ultrasonography and CT may help to come to the diagnosis, the typical findings the spleen being not in normal position.

Treatment of the ectopic spleen is operative and consists of either splenopexy (if not infarcted) or splenectomy.

Case Report

A 41 years female, first visited the hospital on 2058-12-10, with complaints of breathlessness & abdominal discomfort. On examination her general condition was fair, she had no anemia, icterus, clubbing or lymphadenopathy. Her vitals were stable.

The Total Count was190000/cmm with Neutrophils = 60%, Lymphocytes = 19%, Monocytes = 03%, Myeloblast = 02%, Myelocytes = 22%, Metamyelocytes = 04%, Platelets = 155000/cmm and Haemoglobin = 8.9 gm%.

The USG of Abdomen & Pelvis was suggestive of Mild Hepatomegaly with Splenomegaly. So she was diagnosed as *chronic myelocitic leukaemia* and managed conservatively by the physician and was discharged with Hydroxyurea 500mg B.D. Patient was on regular follow up as an out patient basis. But the patient revisited after 4 years (on 062/1/22) with complaints of increasing abdominal mass, increase frequency of micturation and abdominal discomfort. On Examination, her general condition was fair, vitals were stable and her Chest and CVS examination revealed no abnormality.

Per abdominal examination revealed *a large abdominal mass occupying almost all the region of abdomen. It was firm, non tender, mobile, non pulsatile with smooth surface, round border and did not move with respiration. It was dull on percussion and had no bruit.*

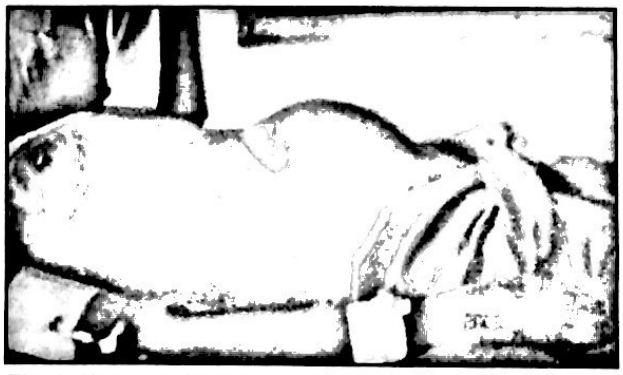

Fig. 1: Showing a large intra- abdominal mass per abdominally.

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This time the Total Count was 134000/cmm with Neutrophils= 67%, Lymphocytes = 02%, Basophils= 05% and Myelocytes=16%. FNAC findings were consistent with *granulocytic sarcoma.*

The Bone Marrow Examination revealed Granulocytic hyperplasia consistent with chronic myelocytic leukaemia.

The subsequent diagnosis was *chronic myelocytic leukamia with massive intra-abdomen mass.*

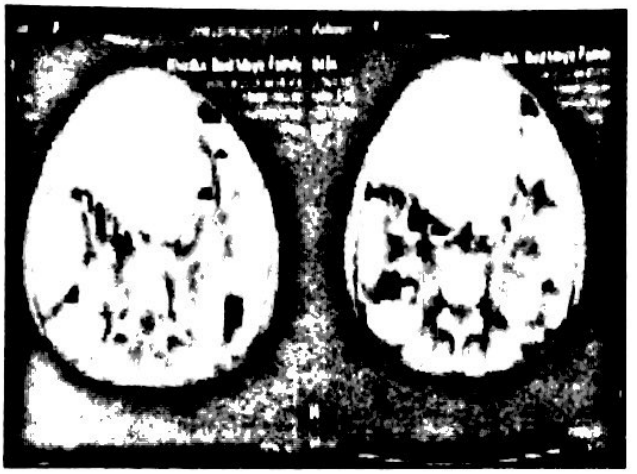

Fig. 2 :

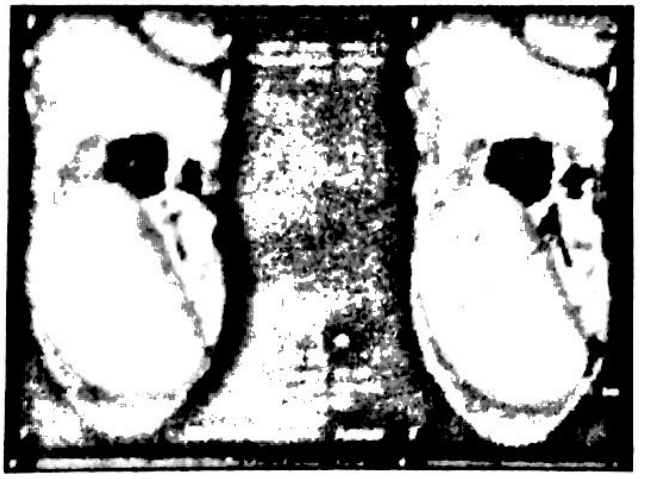

Fig. 3 :

Fig. 2 & 3 Shwoing A massive intra-abdominal mass extending up-to the pelvis.

The case was referred to oncologist and then for surgical consultation. Repeat investigations revealed huge hepatospleenomegaly (USG of Abdomen & Pelvis & CECT) and our clinical suspicion was *a large ectopic spleen.*

Exploratory laprotomy done on 2062-01-26 and patient received Pneumovac and heparin prophylaxis.

Operative Findings

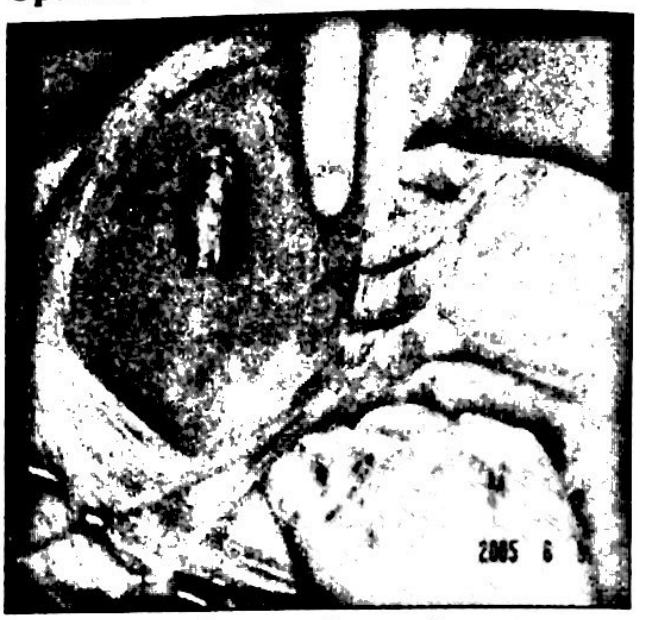

Fig. 4 : Showing rudimentary ligaments

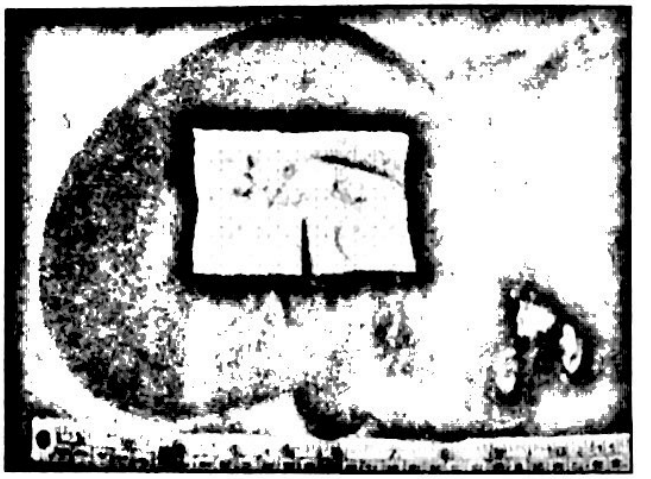

Fig. 5 : Splenectomized specimen.

Grossly enlarged ectopic spleen involving whole abdomen and extending into the pelvic cavity. Splenic ligaments were rudimentary. Hepatomegaly was also associated.

The post operative period was followed with soakage of the dressing due to heparin over dose which was managed with Inj. Protamine sulphate 20mg iv & FFP of IV units. The rest of the post operative period was uneventful. The heparin was subsequently changed into warfarin.

The patient was discharged with Penicillin V 500mg OD, Hydroxurea 500mg BD & is regular follow up at our hospital

Introduction

Anatomy

The spleen is situated in the left upper quadrant tucked under the left dome of the diaphragm and overlain by the lower 9-11th left ribs such that it is impalpable and enlarges 2-3 times before it become palpable on examination. It is overlapped anteriorly by the fundus of the stomach with its long axis lying along the line of the 10th rib. The normal dimension of the spleen is 13x9x3cm, although size even in healthy individual varies considerably. It has an anterior and posterior extremity (pole), superior and inferior borders and two surfaces: visceral facing antero-inferiorly and convex postero-superior (diaphragmatic). The visceral surface is in contact with the stomach, the tail of pancreas, left kidney (and adrenal) and splenic flexure of the colon, all of which cast impression on this surface. The spleen is attached to the diaphragm and retroperitoneal organs by peritoneal folds: splenocolic, diaphragmatic and linorenal. The attachment of the fundus of the stomach is via the gastrosplenic section of the greater omentum that contains the short gastric vessels.

The spleen varies in shape and three morphological types are described: crescentic (hilum extends from prominent superior to inferior poles), rhomboid (hilum forms a wide U) and triangular (with triangular hilum).

It consists of red pulp (80%) made up of sinuses and sinusoids and cellular cords containing macrophages, and white pulp (20%) consisting lymphoid tissue. The red pulp is concerned with maturation and removal of the damaged and senescent red cells, whereas the white pulp forms a major component of the cellular immune surveillance and the protection of the body.

Splenic Funcition

The spleen has important hematopoitic function during early fetal life with both red and white blood cell production. By the 5th month of gestation, the bone marrow assumes the predominant role in hematopoiesis, and normally there is no significant hematopoitic function left in the spleen. Under certain pathological conditions, however such as myelodysplasia, the spleen can reacquire its function. Removal of the spleen does not usually result in anemia or leucopenia in an otherwise healthy person. Although the hematopoitic function is usually lost during fetal development, the spleen continues t o function as sophisticated filter because of the unique circulatory system and lymphoid organization, and it has blood cell monitoring and management function as well as important immune functions throughout life.

The most important function of the spleen is its mechanical filtration (in normal subject and in patients with disease). It also play part in the maintenance of normal immune function and host defense against certain type of infectious disease specially streptococci ,hemophilus and diplococci (encapsulated).

The spleen is a major site of production for the opsonins properdin and tuftsin.

Late Morbitidy After Splenectomy

Postsplenectomy thrombocytosis may be associated with both hemorrhagic and thromboembolic phenomena. This occurs particularly in patients with myeloproliferative disorders such as CML, agnogenic myeloid dysplasia, essential thrombocytosis, and polycythemia vera. Thrombosis in the mesenteric, portal, and renal veins may be a life-threatening sequela of Postsplenectomy thrombocytosis. The lifelong risk of deep venous thrombosis and pulmonary embolism has not been well defined but may be significant. . Pimp et al showed pulmonary embolism as the major or contributory cause of death more often in the splenectomy group (35.6%) than in the control group (9.7%).

OPSI is among the more devastating sequela of asplenia and is the most common fatal late complication of splenectomy. The exact incidence

of OPSI has been difficult to determine. The incidence of infection in post-splenectomy patients is likely to be under-reported. One consistent observation is that the risk for OPSI is greater after splenectomy for malignancy or hematological disease than for trauma. The risk also appears to be greater in young children (<4 years of age). The rise for fatal OPSI is estimated to be 1 per 300 to 350 patient-years follow-up for children and 1 per 800 to 1000 patient-years follow-up for adults. The incidence of nonfatal infection and sepsis is likely to be significantly greater.

S.Pneumoniae is the most frequently involved organism in OPSI and is estimated to be responsible for 50% and 90% of cases. Other organisms involved in OPSI include

Hemophilus influenza, Neisseria meningitides. Streptococcus species and other than pneumococcal species are salmonella species and Capnocytophaga canimorsus (implicated in OPSI as a sequela of dog bites).

Prophylactic Treatment of Splenectomized Patients

The spleen is important for generating responses to thymus-independent antigens. In elective procedures, immunization should be administered before splenectomy whenever possible; the Advisory Committee on Immunization Particles has recommended that the immunization precede splenectomy by at least 2 weeks.

Presplenectomy immunization is not possible in cases of splenic trauma. High-risk patients without spleen should be considered for revaccination if they received the earlier 1+valent vaccine rather than the more current 23-valent preparation or if more than 3 to 6 years have elapsed since primary immunization. Simultaneous immunization with H.influenzae type b, meningococcal serogroup C, and polyvalent pneumococcal vaccine is both immunogenic and well tolerated. Unfortunately, rare cases of OPSI have been reported in vaccinated patients.

Penicillin prophylaxis is commonly practiced in children during the first few years after splenectomy and some authorities have advocated this form of prophylaxis in adults, although data showing the efficacy of this treatment are lacking. OPSI has been reported in both adults and children taking prophylactic penicillin, despite penicillin-sensitive pneumococcal infection. Available data do not support the practice of long-term penicillin prophylaxis in asplenic patients.

Discussion

It is known that the spleen represents one fourth of the total lymphatic mass and it serves as a biological filter for the clearance of bacteria. The spleen is essential for rapid antibody production after challenge with blood-borne particulate antigen in the absence of preexisting antibodies. In addition the spleen appears to be the site of production of a nonspecific leucophilic immunoglobin tuftsin that increases the phagocytic activity of polymorphonuclear leucocytes. Thus the spleen has important functions and its removal is not to be taken lightly. Surgical splenectomy carries a significant postoperative morbidity and a long-term risk of overwhelming infection in 1- 2% of patients. Splenectomy should be avoided whenever possible in favour of conservative medical and surgical approaches. But there are definite indications, in our case it was massive ectopic spleen with 3.2 kg in infracted condition and to perform splenectomy in a diseased condition (Malignancy) is always a challenging one.

But in the experienced hand ectopic spleen can be diagnosed early and the complication can be avoided before hand

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