Case Report

Unusual presentation of Splenic Vein Thrombosis and Infarct as Left Pleural Effusion

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Abstract: -
Pleural effusions are common in clinical practice, commonly due to parenchymal or pleural causes. Splenic vein thrombosis presenting as left pleural effusion is extremely rare. We report a case of a recently diagnosed case of polycythaemia Vera who presented with left sided pleural effusion, which was secondary to splenic infarction due to splenic vein thrombosis.

Background
A left-sided pleural effusion is an infrequent clinical occurrence compared with bilateral or right-sided effusions. Koehler² et al reported the association of left –sided pleural effusions with sub capsular splenic hematoma in 1980. Warren¹ et al published a case report of left-sided pleural effusion that resulted from splenic vein thrombosis. Thromboembolic complications of varying severity occur in about 20 to 30% of the polycythemia vera patients, either at diagnosis or in the course of the disease. We report a similar case of a recently diagnosed case of polycythemia vera who presented with left sided pleural effusion which was secondary to splenic infarction due to splenic vein thrombosis.

Case Report
A 30 year old lady presented in January 2012 with history of mass per abdomen and abdominal discomfort. Investigation revealed splenomegaly and elevated haemoglobin (19.5gm/dl),leucocytosis(14,500/cu mm) and a platelet count of 5.5L/cu mm. Bone marrow studies showed a hyper cellular marrow with marked proliferation of megakaryocytes, increased granulopoiesis and erythropoiesis. There
was no significant reticulin fibrosis. Qualitative RT-PCR for bcr-abl was negative. Jak-2 mutation study was positive. Serum erythropoietin level was in normal range. She was put on low dose aspirin and asked to undergo regular phlebotomies and follow up. Her Haematocrit on discharge was 44. She was however not regular in follow up. She presented in March 2012 with left sided pain abdomen and slight breathlessness. Examination revealed decreased breath sounds in left infrascapular and infra axillary areas with reduced vocal resonance. Abdomen was soft. Rest of the examination was normal. Chest X-ray revealed a left sided pleural effusion. CBC showed a haematocrit of 48, TLC was 12,700/cu mm and platelet count of 7.59L/cumm. CT chest and abdomen revealed moderate left sided pleural effusion with basal atelectasis; large irregular wedge shaped non enhancing hypo dense areas in splenic parenchyma extending from periphery to hilum suggestive of splenic infarcts, splenomegaly, minimum ascites, chronic splenic vein thrombosis and portal vein thrombosis with cavernoma. The other parameters were normal.

Pleural fluid was exudative with total protein of 3.8g/dl, cell count of 2800/cu mm, with plenty of RBCs, neutrophils, few lymphocytes and mesothelial cells. There were no atypical cells. Bacterial, fungal and AFB cultures were negative. LDH was 60 IU/L. She was given symptomatic treatment with hydration, analgesics and a course of antibiotics. Aspirin was continued and anticoagulation was started with warfarin. She was started on cyto reductive agent- Hydroxyurea. Therapeutic thoracocentesis was done. However there was repeated recollection of fluid within a few days and she continued to be symptomatic. Hence we decided to go ahead with splenectomy. The spleen was enlarged intraoperatively and histopathology revealed red pulp hyperplasia, infarction and haemorrhage.

She is on regular follow up now and there has been no recurrence of the pleural effusion nine months after the splenectomy.

Discussion
Pleural effusions could be multifactorial. The aetiology is usually of parenchymal or pleural origin. Sub phrenic causes for pleural effusions are rare and usually go unsuspected. There are only a few anecdotal reports of splenic pathology leading to left sided pleural effusions, which have completely resolved after splenectomy.

Polycythaemia is a clonal haemopoietic chronic myeloproliferative disorder characterized by an excessive production of all the marrow elements i.e., the nucleated red cells, the granulocytes and the megakaryocytes. The course of polycythaemia is complicated by haemorrhagic and/or thromboembolic complications of varying severity in about 20 to 30% of the patients. The median age of onset is 55-60 years whereas our patient was quite young.

The pathogenesis of thrombosis in myeloproliferative neoplasms is multifactorial and the relative role of these abnormalities, as compared with that of other individual and environmental factors, is controversial. Quantitative and qualitative red blood cell, platelet, and leukocyte abnormalities are likely to play a key-role in myeloproliferative neoplasm thrombophilia. High shear stress of the vessel wall, due to blood hyper viscosity, accounts for chronic endothelial dysfunction and platelet
and leukocyte activation. The increase of the thrombotic risk observed at progressively higher haematocrit values parallels blood viscosity, although also biochemical changes in the cell membrane and content could contribute to rheological abnormalities. Platelets and endothelial cells play a pivotal role in regulating blood flow; both cells might contribute to determine a prothrombotic microenvironment in myeloproliferative neoplasm patients by producing more soluble selectins and less nitric oxide, likely as a consequence of inflammation. Leucocytosis has been shown to represent an independent risk factor for thrombosis.

![Diagram](image)

Pleural effusion secondary to splenic vein thrombosis and infarction is very rare with very few reported cases. Pleural effusion is commonly due to diseases of the pleura, the lung parenchyma, or vasculature, and rarely extra pulmonary disorders. Among the many possible aetiologies of pleural effusion in adults, the most common are pneumonia, heart failure, malignancy, tuberculosis, and pulmonary embolism, collagen vascular disorders and hypoalbuminemic states (cirrhosis, nephrotic syndrome, etc).

An understanding of the lymphatic drainage of the pleura is important. The costal pleura drain into internal mammary and intercostal lymphatic system. The visceral pleura drains into mediastinal lymph nodes. The lymph passes sequentially through the inferior and superior tracheobronchial nodes, paratracheal nodes, bronchomediastinal trunk, and finally to the right lymphatic or thoracic duct. The Thoracic duct drains all of the body and limbs below the respiratory diaphragm; the left side of the chest, left upper limb and the left side of the head and neck above the diaphragm. There are also unidirectional trans diaphragmatic vessels that descend and drain the posterior thorax receiving tributaries from the diaphragm and splenic mesentery.

The postulated mechanism of pleural effusion proposed by Warren et al in such cases include:

1. Direct compression of the posterior lymphatics by the enlarged infarcted spleen
2. Filtration of the haemorrhagic pleural fluid into the pleural space due to increased permeability caused by perisplenic inflammation. Leung et al have reported pleural effusion associated with urinary tract obstruction due compression of posterior lymphatics. Miller and Talman have described the relationship of sub phrenic inflammation and increased filtration of peritoneal fluid into pleural space.

There was no other known cause for pleural effusion in this case and the effusion cleared after splenectomy supporting a cause and effect relationship. Hence in patients with recurrent or persistent left pleural effusions, sub diaphragmatic causes of effusions should also be considered and systematically investigated.

Note: Written consent of the patient was taken for publication of these details.

References:


Figures
Fig 1: X-ray at presentation showing Left pleural effusion

Fig 2: X-ray shows significant resolution after thoracocentesis but recurred

Fig 3: X-ray on follow up after 9 months