

DEFINITION

1. **Myelofibrosis** is a neoplastic myeloproliferative disorder in which there occurs a progressive patchy fibrous replacement of the bone marrow due to increased fibroblastic activity in the marrow. This fibrosis is a reaction to the disease process, not part of the neoplastic disorder.

CLINICAL MANIFESTATIONS

2. The condition occurs in elderly people, and usually has an insidious onset. The spleen is enlarged and abdominal pain due to splenic infarction occurs. The liver may also be enlarged, with both organs being the site of myeloid metaplasia. Bone marrow sampling establishes the diagnosis.
3. The peripheral blood picture is one of anaemia, marked polychromasia, anisocytosis, and poikilocytosis. There may be folate deficiency and macrocytosis. Nucleated red cells and immature granulocytes give a leuco-erythroblastic picture. At the time of diagnosis the granulocyte count is variable but the platelet count is usually increased. As the disease progresses all formed elements tend to decrease. The neutrophil alkaline phosphatase and serum uric acid levels are raised.

AETIOLOGY

4. **Myelofibrosis** may be **primary** or **secondary**, in the latter case being associated with other neoplastic disorders of the bone marrow, metabolic changes involving vitamin D or its metabolites and a variety of other conditions which provoke a fibrous reaction by an unknown mechanism.
5. Despite extensive study, the cause of **primary myelofibrosis** is unknown but it seems that the abnormal haemopoietic elements result from neoplastic proliferation of an abnormal stem cell population. Secondary fibrosis of the bone marrow and myeloid metaplasia of liver, spleen, and lymph nodes thus occur simultaneously. The aetiology of **secondary myelofibrosis** is that of the underlying condition.

CONCLUSION

6. **Myelofibrosis** is a neoplastic disorder of the bone marrow, the aetiology of which is unknown.

REFERENCE

Weatherall D J. Myelosclerosis. In: (Eds) Weatherall D J, Ledingham J G G, Warrell D A. Oxford Textbook of Medicine. 2nd Ed. 1987. Oxford. Oxford University Press. p9.40–19.46.

December 1992