Primary myelofibrosis is a rare condition where myelofibrosis leads to bone marrow failure. It is often idiopathic, but a JAK-2 mutation and cases linked to benzene exposure are described.

A 38-year-old Hispanic male presented with a 2-week history of fever, chills, and a worsening generalized maculopapular rash, associated with amoxicillin and ibuprofen given after dental work. He had dry cough, sore throat, coryza, and generalized malaise. He works in an automotive repair shop. Endocarditis was considered given his history of dental work, but imaging studies showed hepatosplenomegaly and lymphadenopathy. Hepatitis panel and HIV tests were negative.

Evaluation included CBC, peripheral blood smear, chromosome analysis, bone marrow aspirate and biopsy, FISH for ABL/BCR rearrangement, flow cytometry, and molecular studies (JAK-2 and CALR mutational studies).

The 2008 WHO criteria for diagnosis of primary myelofibrosis requires all three major criteria and at least two minor criteria. The major criteria demonstrated: 1) megakaryocytic atypia and proliferation and 2) exclusion of other myeloproliferative disorders. He also fulfills two minor criteria, consisting of anemia and splenomegaly. An MPL mutation test was recommended to further support the diagnosis, but had not yet been performed at the time of this writing.