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# Neurofibromatosis with chronic myeloid leukemia in an elderly male: An unusual association

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## Abstract

Neurofibromatosis is a genetic disorder of neural crest-derived cells that predominantly affect growth and development of neural tissues. We report a case of 64-year-old patient, had several soft tissue cutaneous nodules (neurofibroma) on the body including the face, head, and neck, extremities and multiple hyperpigmented macules on trunk and back (Café-au-lait pigmentation), who was accidentally diagnosed as chronic myeloid leukemia, on routine investigation for surgical management. He did not have any systemic manifestation either of diseases.

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## Full Text

### INTRODUCTION

Neurofibromatosis 1 (NF1) is an autosomal dominant progressive neurocuteneous disorder caused by a wide variety of mutations that affect the NF1 gene located at 17q11.2 chromosome. NF1 has one of the foremost rates of spontaneous mutation among genetic disorder in humans. The association of NF1 and chronic myeloid leukemia (CML) in adult age group is rarely encountered in clinical practice. Bader and Miller. [1] Reported an increased magnitude of nonlymphocytic leukemia among children with NF1 and leukemia. The overall risk of cancer

malignancies in NF1 is 2.7-fold increased then general population. Stiller et al. [2] Conducted a population-based study, found an increased relative risk of chronic myelomonocytic leukemia, acute lymphoblastic leukemia, and non-Hodgkin's lymphoma in the NF1 population. The association between germ-line mutations of NF1 and hematologic malignancies has been established. [3]

## CASE REPORT

A 64-year-old man presented with complaints of both leg bones fracture due to an accidental injury. The patient was diagnosed as nonfamilial NF1 40 years back. The routine blood examination revealed >2 lacs white blood cell counts. On peripheral blood film examination, shows full spectrum of cells ranging from myeloblasts to mature neutrophils with predominant immature myeloid cells (metamyelocytes and myelocytes) and also the numbers of basophils and eosinophils increased [Figure 1]. Physical examination was remarkable for cutaneous circumscribed nodules which appeared soft to the touch, present over face, head, and neck, extremities and back along with multiple hyperpigmented macules on trunk and back (Café-au-lait pigmentation) [Figure 2]. He had no evidence of splenomegaly. The bone marrow picture revealed CML in blast phase. A biopsy of the tissue fragment from the nodules confirmed the presence of neurofibromatosis. The Bone marrow cytology that included cytogenetic study confirmed the presence of CML with a positive Philadelphia chromosome. The BCR-ABL was detected using real-time polymerase chain reaction (RT-PCR) assay; the signals for BCR-ABL were detected in leucocytes of the specimens. The patient has two children with no history of NF in either of them. Prescription of 400 mg oral imatinib mesylate daily yielded complete hematological remission at end of 2 months and partial cytogenetic response at end of 6 months of treatment. At 12 months follow-up, complete cytogenetic response was seen by RT-PCR. This case illustrates a sporadic association between CML and NF in an adult male.{Figure 1}{Figure 2}

## DISCUSSION

Neurofibromatosis 1 is a tumor suppressor gene that encodes the neurofibromin, a negative regulator of proto-oncogene Ras, so loss of neurofibromin promotes Ras activity leading to downstream signaling and increased uncontrolled cell growth. [4] The uncontrolled growth predisposes to both cancerous and noncancerous disorders. The risk of malignancies in patients with NF1 is well-established, and increases with age, the majority of cancers reported are nonhematologic neoplasms. [5] NF1 is a common inherited disorder with a high number of potentially associated pathologies and malignancies, but the association of leukemia and NF1 has rarely been demonstrated in adults. [6] A study by Olayemi et al. Reported a 35-year-old female with NF, who developed CML at a 35-year-old female. [7] Sartor et al. also reported a case of 65-year-old patient affected by NF1 and acute myeloid leukemia. [8] Imatinib mesylate is the current standard first-line therapy for all phases of CML. We conclude that the association of NF1 and CML is a sporadic one, and the treating oncologist should be aware of this rare association.

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