Chronic neutrophilic leukaemia (CNL) is a rare BCR/ABL negative myeloproliferative disorder characterised by persistent neutrophilia and splenomegaly. Most patients with CNL have a poor prognosis, with a mean survival time of 14.7 months. To date, only 129 cases of CNL have been reported in the literature. The term “true” CNL recently introduced by Reilly highlights the need for more experience with further CNL cases to improve the diagnostic criteria. Therefore, we present our findings in a large group of 14 new cases of CNL, consisting of eight women and six men (mean age, 64.7 years).

**Aims:** To report the findings from a large group of 14 new cases of CNL, consisting of eight women and six men (mean age, 64.7 years).

**Methods:** A review of the 14 new cases of CNL and the investigation of BCR/ABL translocations in these patients.

**Results:** Three quarters of the patients died within two years after diagnosis, mostly as a result of severe cerebral haemorrhage. Two younger patients were successfully treated with allogeneic bone marrow transplantation or interferon, which resulted in haematological remission for years.

**Conclusion:** CNL is a rare myeloproliferative disease mostly taking a fatal clinical course, despite the presence of mature neutrophils as leukaemic cells in the blood. Thus, it is important to recognise CNL to develop appropriate therapeutic strategies for affected patients.

**DISCUSSION**

The main diagnostic criteria of CNL are chronic neutrophilia in the blood, expansion of neutrophilic granulopoiesis in the bone marrow, and splenomegaly in the absence of any form of BCR/ABL translocation or leukaemic reaction. These criteria were sufficiently fulfilled in our 14 patients with CNL. The spectrum of fatal complications we saw in our patients was very similar to that described in the literature. The haemorrhagic diathesis seen in patients with CNL may be the result of thrombocytopenia and thrombocyte dysfunction, or it may be caused by leukaemic infiltration of vascular walls. With rare exceptions, CNL is a disease of older adults.

At the time of diagnosis, 88% of the patients with CNL in the literature were older than 50 years. The sex distribution in CNL is nearly equal. There is doubt about whether all of the CNL cases in the literature represent true CNL. Some authors have suggested that CNL is nearly equal. There is doubt about whether all of the CNL cases in the literature represent true CNL. Some authors have suggested that CNL is nearly equal. There is doubt about whether all of the CNL cases in the literature represent true CNL.

**Abbreviations:** CML, chronic myeloid leukaemia; CNL, chronic neutrophilic leukaemia; NAP, neutrophil alkaline phosphatase.
that those cases of CNL that occurred in association with plasma cell dyscrasias like myeloma were in fact neutrophilic reactions. Moreover, it was suggested that cases of CNL showing dysplastic features would be better classified as a myelodysplastic entity. Thus, reviewing the data of all CNL cases in the literature, Reilly defined a group of 33 cases of true CNL, including one unpublished case of his own. This group of 33 selected patients with CNL also showed a high mean age (62.5 years) and short survival times (mean survival, 30 months), but had a 2 : 1 male to female ratio. The term true CNL used by Reilly reveals the need for an even more precise definition of CNL as an entity. Thus, the diagnostic criteria of CNL should be applied in a strict manner, especially for the conditions mentioned above.

"To date, because of the rarity of the disease, no therapeutic standard has been determined in chronic neutrophilic leukaemia"
Take home messages

- Chronic neutrophilic leukaemia (CNL) is a rare myeloproliferative disease, mainly found in elderly patients
- This disease has a mostly fatal outcome—three quarters of our patients died within two years of diagnosis, mainly as a result of severe cerebral haemorrhage
- Two younger patients were successfully treated with autologous bone marrow transplantation or interferon, which resulted in haematological remission for years
- Thus, it is important to recognise CNL and to develop appropriate therapeutic strategies for affected patients

agents, such as hydroxyurea, may temporarily control leucocytosis and splenomegaly, and the use of interferon α may induce long standing clinical remission. So far, autologous bone marrow transplantation represents the only treatment modality with curative potential. We conclude that it is important to recognise CNL as a rare, but distinct, disease entity different from CML, and in particular to distinguish CNL from leukaemoid reactions, because patients with CNL generally have a poor prognosis. To gain a better understanding of the nature of true CNL the reporting of new cases must be encouraged.

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