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# Familial neurofibromatosis type I and adult acute lymphocytic leukemia

Sir,

Individuals with neurofibromatosis type I (NF1; von Recklinghausen's disease) are predisposed to certain cancers. Children are at increased risk of developing benign and malignant solid tumors (mostly neural tumors) as well as hematologic malignancies including juvenile myelomonocytic leukemia (a rare hematologic malignancy that affects patients less than 4 years of age and that is sometimes associated with monosomy 7), the monosomy 7 myelodysplastic syndrome and acute myelogenous leukemia.<sup>1</sup> The risk of young children with NFI developing a malignant myeloid disorder is 200 to 500 greater than the normal risk<sup>2</sup> and several lines of evidence support the notion that the loss or mutation of the NF1 gene (a tumor suppressor gene) deregulates the Ras pathway which is responsible for the leukemogenesis in these children.<sup>3</sup> The relative risks for non-Hodgkin's lymphoma and acute lymphocytic leukemia (ALL) were increased in one series<sup>2</sup> but the risk of developing ALL was not increased in two other reports.<sup>4,5</sup>

In contrast to the situation in childhood, the association between NF1 and malignant blood disorders has not been demonstrated in adulthood.<sup>3,6,7</sup> A Medline<sup>®</sup> search of reports from the last 10 years uncovered only 2 adult patients with NF1 who developed an acute myelogenous leukemia<sup>6,8</sup> and none an ALL

It is, therefore, of considerable interest that we have seen 2 cases of adult-ALL, diagnosed over the last 12 months, in patients with familial NF1. This represents a 1.1% incidence in 176 ALL in patients over the age of 14 years seen in our Service during the last 25 years.

The diagnosis of NF1 is mainly clinical (*café-au-lait* spots, freckling, cutaneous neurofibromas, lentigo, Lisch's nodules). Our two patients had these signs and several relatives with the disease (Table 1). Case #1 was a 19 year-old male with a familial history of NF1 (mother and the only sister affected) and case #2 a 31 year-old woman with NF1 extending through 4 generations (grandfather, mother, 2 uncles, 1 aunt and her only 14-year-old son affected). Both patients had the common ALL cell phenotype, a normal karyo-type and very aggressive clinical evolution.

#### Table 1. Clinical characteristics of the patients.

	Case #1	Case #2
Age at diagnosis of ALL/sex	19/M	31/F
Familial history of neurofibromatosis (affected relatives)	Mother Sister Uncles (2) Aunt Son	Grandfather Mother
Clinical features of neurofibromatosis: >6 café-au-lait spots Cutaneous neurofibromas Freckles Other abnormalities	+ + +	+ + -
ALL phenotype	Common	Common
Karyotype	Normal	Normal

Because NF1 in one of the most common autosomal dominant disorders (with an incidence of 1:3000 neonates), one must be aware that the association observed could easily occur by chance. Although case reports are not appropiate for causation assessment, they have the potential to generate new knowledge by stimulating reporting of additional single cases or the development of more traditional epidemiologic surveys that may lead to a precise estimate of risk. For additional information about the molecular pathoegensis of acute lymphoblastic leukemia, the reader is referred to recent papers in this journal.<sup>9,10</sup>

Thus, besides the well known association between NF1 and myeloid disorders in children, our two case reports reveal a possible causal relation between familial NF1 and adult onset ALL.

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### Key words

Neurofibromatosis, ALL

### Correspondence

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## Central nervous system involvement in acute promyelocytic leukemia. A description of two cases and review of the literature

## Sir,

Extramedullary involvement is infrequent in acute promyelocytic leukemia (APL), but it has been suggested that its incidence might be increasing. Two patients with APL and central nervous system (CNS) involvement are described and the possible relationship between this complication and new treatment approaches of APL are discussed.

*Case #1.* A 45-year-old woman was diagnosed with APL. Cytogenetic studies revealed a complex karyotype in bone marrow (46,XX,r(9) t(17;15;10) (q11; q22;q24) and a bcr-1 pattern of the PML-RAR $\alpha$  fusion protein. A molecular complete remission (CR) was achieved within the first month from starting ATRA and standard chemotherapy. Five days after administration of a consolidation course she presented with a third cranial nerve palsy and examination of the cerebrospinal fluid revealed the presence of promyelocytes in which the PML-RAR $\alpha$  rearrangement was detected. She was given ATRA plus intrathecal chemotherapy without achieving a response and died shortly thereafter from progressive disease.

*Case #2.* A 45-year-old man was diagnosed with APL and a bcr-1 pattern of the PML-RAR $\alpha$  fusion protein was demonstrated in peripheral blood. CR was achieved after treatment with ATRA and standard chemotherapy. Twenty months later he presented with a bone marrow relapse. After attaining a second CR he

Bernengo     1975     82/M     No     Cutaneous     Om     NA     Death (PD)       Belasco     1978     9m/M     No     Soft lissue     Om     NA     CR       Nihei     1984     55/M     No     Mediasinum     Om     NA     Death (PD)       Kubonishi     1984     25/M     No     Intracencral mass, CSF     Om     NA     Death (PD)       Kanakura     1989     59/F     No     Cutaneous     36/m     NA     BM relapse       Zuiable     1989     31/M     No     CR (zatrous and CNS     NA     NA     Death (PD)       Ajarim     1990     21/M     No     Catrous and CNS     NA     NA     Death (PD)       Brown     1992     37/F     No     Optic nerve (also BM)     22m     Yes     CR (NA)       Urisar     1994     63/F     Yes     Cataneous And CNS     NA     NA     Death (PD)       Brown     1994     37/M     Yes     Cataneous And CNS     10m     Yes	Author	Year	A/S	ATRA*	Extramedullary site	Interval	Cyto	Outcome
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Kubonishi     1984     23/M     No     Mediasinum     Om     NA     Death (P)       Kanakura     1987     44/F     No     Infracerebral mass, CSF     Om     NA     CR (23m)       Baer     1989     31/M     No     SMC (extradural mass)     Om     No     BM relapse       Zuiable     1989     13/M     No     Mediastinum (also BM)     Om     No     BM relapse       Nazi     1991     26/M     No     Outaneous and CNS     NA     NA     Death (PD)       Brown     1992     37/F     No     Optic nerve (also BM)     22m     Yes     CR (NA)       Iongare     1993     19/M     NA     Cutaneous     10m     Yes     CR (NA)       Iongare     1993     37/M     Yes     Cutaneous CSF     13m     Yes     CR (NA)       Girait     1994     37/M     Yes     Cutaneous CSF     13m     Yes     CR (NA)       Girait     1994     37/M     Yes     Cutaneous CSF     13m <td< td=""><td>Nihei</td><td>1984</td><td>55/M</td><td>No</td><td>Mediastinum and muscle</td><td>4m</td><td>NA</td><td>Death (PD)</td></td<>	Nihei	1984	55/M	No	Mediastinum and muscle	4m	NA	Death (PD)
Kanakura     1987     44/F     No     Intracerebral mass, CSF     Om     NA     CR (23m)       Baer     1989     59/F     No     Cutaneous     36m     NA     BM relapse       Zuiable     1989     31/M     No     SNC (extradural mass)     Om     No     BM relapse       Rush     1990     13m/M     No     Mandibular     22m     No     CR (16m)       Ajarim     1990     21/M     No     Outaneous and CNS     NA     NA     Dattereous and CNS     NA     NA     Dattereous and CNS     NA     NA     Dattereous and CNS     NA     NA <td< td=""><td>Kubonishi</td><td>1984</td><td>23/M</td><td>No</td><td>Mediastinum</td><td>Om</td><td>NA</td><td>Death (PD)</td></td<>	Kubonishi	1984	23/M	No	Mediastinum	Om	NA	Death (PD)
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Itsi     1995     27/W     No     Epidular     Offi     Tes     CR (1411)       Hazneradoglu 1995     19/M     No     Gingiva     15m     NA     Death (PD)       Bekassy     1995     24/F     NA     Outaneous     NA     NA     Response       Bekassy     1995     3/F     NA     Spinal     NA     NA     NA     Death (PD)       Bekassy     1995     26/M     NA     Testicle&spinal     NA     NA     Death (PD)       Bekassy     1995     3/F     No     Cutaneous     18m     Yes     Response       Selleri     1996     31/F     No     Cutaneous (also BM)     6m     Yes     Death (PD)       Wiernik     1996     5/F     Yes     Gingiva     21m     Yes     Death (PD)       Matrinelli     1997     2/M     No     Cutaneous (also BM)     6m     Yes     Death (PD)       Matrinelli     1997     2/M     Yes     CNS     15m     Yes     Death (PD) </td <td>Gildil</td> <td>1994 100E</td> <td>4//F</td> <td>162</td> <td>Cuidieous</td> <td>0m</td> <td>INA Vee</td> <td>CD (14m)</td>	Gildil	1994 100E	4//F	162	Cuidieous	0m	INA Vee	CD (14m)
Hazita adugut 1995197/MNACutaneousNANAResponseBekassy199524/FNAQutaneousNANAResponseBekassy19953/FNASpinalNANADeath (PD)Bekassy19953/FNASpinalNANADeath (PD)Bekassy19953/FNoSubcutaneous&CNS6mYesResponseSelleri199631/FNoCutaneous18mYesCR (25m)Wiemik19965/FYesGingiva21mYesDeath (PD)Wiemik19965/FNoCutaneous (also BM)6mYesDeath (PD)Chan199625/FNoCutaneous (also BM)6mYesDeath (PD)Martinelli199742/MNoSpleen0mYesDeath (VD)Martinelli199742/MNoBone (L4)0mYesDeath (VD)Evans199722/MYesCNS(CSF+)NAYesDeath (PD)Molero199740/MYesCutaneous (also BM)7mYesDeath (PD)Ueda199728/FNoCutaneous (also BM)7mNAPDLiso199724/MYesColonNAYesDeath (PD)Ueda199724/MYesColonNAYesDeath (PD)Liso199847/MNoCutaneous11m	1051	1995 1005	2//IVI 10/M	No.	Epidural	UIII 1Em	Tes	UK (1411) Death (DD)
Beckassy199524/FNACutaneousNANANADeath (PD)Bekassy19953/FNASpinalNANADeath (PD)Bekassy199526/MNATesticle&spinalNANADeath (PD)Bekassy199526/MNATesticle&spinalNANADeath (DD)Lederman199546/FNoSuboutaneous&CNS6mYesResponseSelleri19965/FYesGingiva21mYesDeath (PD)Wiemik19965/FYesGingiva21mYesDeath (PD)Mernik19965/FNoCutaneous (also BM)6mYesDeath (PD)Chen199674/MNoSpleen0mYesDeath (PD)Marinelli199742/MNoBone (L4)0mYesDeath (PD)Marinelli199723/MYesCNS15mYesDeath (PD)Evans199723/MYesCNS(CSF+)13mYesDeath (PD)Molero199740/MYesCutaneous (also BM)7mNAPDLiso199723/MYesCutaneous2-3mYesDeath (PD)Maloisel199723/MNoCutaneous2-3mYesDeath (PD)Liso199847/MNoCutaneous14mYesDeath (PD)Liso199822/FNoCuta	Hazner auogiu	1993 1005	19/IVI 24/E	INO NA	Gingiva		INA NA	Dealin (PD)
Bekassy1995 $3/F$ NASpiralNANANADeath (rU)Bekassy1995 $26/M$ NATesticle&spinalNANADeath (rU)Lederman1995 $46/F$ NoSuboutaneous&CNS6mYesResponseSelleri1996 $31/F$ NoCutaneous18mYesCR (25m)Wiemik1996 $5/F$ YesGingiva21mYesDeath (PD)Wiemik1996 $5/F$ YesGingiva21mYesDeath (PD)Wiemik1996 $25/F$ NoCutaneous (also BM)6mYesDeath (PD)Chen1996 $25/F$ NoCutaneous (also BM)6mYesDeath (PD)Martinelli1997 $42/M$ NoBone ((4)0mYesDeath (PD)Evans1997 $23/M$ YesCNS15mYesDeath (PD)Evans1997 $40/H$ YesCNS(CSF+)NAYesDeath (PD)Molero1997 $40/M$ YesCutaneous (also BM)7mYesDeath (PD)Ueda1997 $40/M$ YesCutaneous (also BM)7mNAPDKishimolo1997 $12/F$ NoCutaneous (also BM)7mNAPDCastagnola1997 $23/M$ NoCNS11mYesDeath (PD)Iaios1998 $4/M$ NoCutaneous $4$ sosNaYesDeath (PD) <td< td=""><td>Bekassy</td><td>1995</td><td>24/1</td><td>INA NA</td><td>Cutaneous</td><td>INA NA</td><td>NA</td><td>Response</td></td<>	Bekassy	1995	24/1	INA NA	Cutaneous	INA NA	NA	Response
Bekasy Bekasy199526/MNALesticle XgnalNANADeath (RXC)Lederman199631/FNoSubcutaneous&CNS6mYesResponseSelleri199631/FNoCutaneous18mYesCR (25m)Wiernik19965/FYesGingiva21mYesDeath (PD)Wiernik19965/FNoCutaneous (also BM)6mYesDeath (PD)Chen199674/MNoSpleen0mYesDeath (PD)Martinelli199742/MNoBone (L4)0mYesBM relapseEvans199723/MYesCNS15mYesDeath (PD)Varans199722/MYesBone (mastoid)6mYesDeath (PD)Varans199740/MYesCNS(CSF+)NAYesDeath (PD)Ueda199740/MYesCutaneous (also BM)7mYesDeath (PD)Ueda199740/MYesCutaneous (also BM)7mYesDeath (PD)Ueda199723/MNoCutaneous (also BM)7mYesDeath (PD)Kishimoto199723/MNoCNS11mYesDeath (PD)Maloisel199723/MNoCutaneous2.3mYesDeath (PD)Isio19988/MNoCutaneous14mYesDeath (PD)Liso19982/F <td>Bekassy</td> <td>1995</td> <td>3/F</td> <td>INA</td> <td>spinal</td> <td>NA</td> <td>NA</td> <td>Death (PD)</td>	Bekassy	1995	3/F	INA	spinal	NA	NA	Death (PD)
Lederman199546/FNoSubcutaneous&UNS6mYesResponseSelleri199631/FNoCutaneous18mYesCR (25m)Wiernik19965/FYesGingiva21mYesDeath (PD)Wiernik199625/FNoCutaneous (also BM)6mYesDeath (PD)Martinelli199742/MNoSpleen0mYesDeath (PD)Martinelli199742/MNoBone (L4)0mYesDeath (PD)Evans199723/MYesCNS15mYesDeath (PD)Evans199722/MYesBone (mastoid)6mYesDeath (PD)Evans199740/HYesCNS(CSF+)NAYesDeath (PD)Ueda199766/MYesCutaneous (also BM)7mYesNADel Rio199728/FNoCutaneous (also BM)7mNAPDKishimoto199728/FNoCutaneous (also BM)7mNAPDMaloisel199724/MYesColonNAYesDeath (PD)Iso19988/MNoCutaneous2-3mYesDeath (PD)Liso199825/FNoCutaneous14mYesDeath (PD)Liso199825/FNoCutaneous14mYesDeath (PD)Liso199830/MYesCutaneous <td< td=""><td>Bekassy</td><td>1995</td><td>26/M</td><td>NA</td><td>lesticle&amp;spinal</td><td>NA</td><td>NA</td><td>Death (toxic)</td></td<>	Bekassy	1995	26/M	NA	lesticle&spinal	NA	NA	Death (toxic)
Selleri     1996     31/F     No     Cutaneous     18m     Yes     CR (2sm)       Wiernik     1996     5/F     Yes     Gingiva     21m     Yes     Death (PD)       Wiernik     1996     5/F     No     Cutaneous (also BM)     6m     Yes     Death (PD)       Chen     1996     74/M     No     Spleen     0m     Yes     Death (PD)       Martinelli     1997     42/M     No     Bone (L4)     0m     Yes     Death (PD)       Evans     1997     22/M     Yes     Bone (mastoid)     6m     Yes     Death (PD)       Molero     1997     22/M     Yes     Bone (mastoid)     6m     Yes     Death (PD)       Molero     1997     40/M     Yes     CNS(CSF+)     13m     Yes     Death (PD)       Molero     1997     24/F     No     Cutaneous (also BM)     7m     NA     PD       Catagona     1997     24/M     Yes     Colon     NA     Yes     Death (PD)	Lederman	1995	46/F	NO	Subcutaneous&CNS	6m	Yes	Response
Wiernik1996 $5/F$ YesGingiva $21m$ YesDeath (PD)Wiernik1996 $5/F$ NoCutaneous (also BM)6mYesDeath (PD)Chen1996 $74/M$ NoSpleen0mYesDeath (PD)Martinelli1997 $42/M$ NoBone (L4)0mYesDeath (PD)Evans1997 $22/M$ YesBone (L4)0mYesDeath (VHD)Evans1997 $22/M$ YesBone (mastoid)6mYesDeath (VHD)Evans1997 $22/M$ YesBone (mastoid)6mYesDeath (VHD)Evans1997 $40/M$ YesCNS(CSF+)NAYesDeath (PD)Molero1997 $40/M$ YesCutaneous (also BM)7mNAPDLishimoto1997 $28/F$ NoCutaneous (also BM)7mNAPDCastagnola1997 $23/M$ NoCutaneous $2-3m$ YesDeath (PD)Maloisel1997 $24/M$ YesColonNAYesDeath (PD)Forrest1998 $34/M$ NoCutaneous14mYesDeath (PD)Liso1998 $25/F$ NoCutaneous14mYesDeath (PD)Liso1998 $30/M$ YesCutaneous12mYesDeath (PD)Liso1998 $30/M$ YesCutaneous12mYesDeath (PD)Liso1998<	Selleri	1996	31/F	No	Cutaneous	18m	Yes	CR (25m)
Wiernik199625/FNoCutaneous (also BM)6mYesDeath (PD)Chen199674/MNoSpleen0mYesDeath (PD)Martinelli199742/MNoBone (L4)0mYesBM relapseEvans199723/MYesCNS15mYesDeath (VD)Evans199723/MYesCNS15mYesDeath (VD)Evans199723/MYesCNS(CSF+)NAYesDeath (PD)Molero199740/MYesCNS(CSF+)13mYesDeath (PD)Ueda199766/MYesCutaneous (also BM)7mNAPDLical199728/FNoCutaneous (also BM)7mNAPDCastagnola199723/MNoCNS2-3mYesDeath (PD)Maloisel199723/MNoCNS11mYesDeath (PD)Liso199834/MYesColonNAYesDeath (PD)Liso199822/FNoCutaneous14mYesDeath (PD)Liso199825/FNoCutaneous12mYesDeath (PD)Liso199830/MYesCutaneous12mYesDeath (PD)Liso19983/MYesCutaneous12mYesDeath (PD)Liso19983/MYesCutaneous12mYesDeath (PD)	Wiernik	1996	5/F	Yes	Gingiva	21m	Yes	Death (PD)
	Wiernik	1996	25/F	No	Cutaneous (also BM)	6m	Yes	Death (PD)
Martinelli     1997     42/M     No     Bone (L4)     Om     Yes     BM relapse       Evans     1997     23/M     Yes     CNS     15m     Yes     Death (GVHD)       Evans     1997     22/M     Yes     Bone (mastoid)     6m     Yes     CR (NA)       Evans     1997     24/F     Yes     CNS(CSF+)     NA     Yes     Death (PD)       Molero     1997     40/M     Yes     Cutaneous (also BM)     7m     Yes     Death (PD)       Ueda     1997     28/F     No     Cutaneous (also BM)     7m     NA     PD       Kishimoto     1997     11/F     Yes     Cutaneous (also BM)     7m     NA     PD       Castagnola     1997     23/M     No     CNS     11m     Yes     Death (PD)       Maloisel     1997     23/M     No     CNS     11m     Yes     Death (PD)       Gastagnola     1997     24/M     Yes     Colon     NA     Yes     Death (PD) <td>Chen</td> <td>1996</td> <td>74/M</td> <td>No</td> <td>Spleen</td> <td>Om</td> <td>Yes</td> <td>Death (PD)</td>	Chen	1996	74/M	No	Spleen	Om	Yes	Death (PD)
Evans     1997     23/M     Yes     CNS     15m     Yes     Death (GVHD)       Evans     1997     22/M     Yes     Bone (mastoid)     6m     Yes     CR (NA)       Evans     1997     49/F     Yes     RNS(CSF+)     NA     Yes     Death (PD)       Molero     1997     40/M     Yes     CNS(CSF+)     NA     Yes     Death (PD)       Ueda     1997     6/M     Yes     Cutaneous (also BM)     7m     NA     PD       Kishimoto     1997     28/F     No     Cutaneous (also BM)     7m     NA     PD       Kishimoto     1997     23/M     No     Cutaneous     2.3m     Yes     Death (PD)       Maloisel     1997     23/M     No     CNS     11m     Yes     Death (PD)       Kishimoto     1997     34/M     Yes     Colon     NA     Yes     Death (PD)       Liso     1998     48/M     No     Lumph node     25m     Yes     Death (PD)	Martinelli	1997	42/M	No	Bone (L4)	0m	Yes	BM relapse
Evans199722/MYesBone (mastoid)6mYesCR (NA)Evans199749/FYesCNS(CSF+)NAYesDeath (PD)Molero199740/MYesCNS(CSF+)13mYesDeath (PD)Ueda199766/MYesCutaneous (also BM)7mYesNADel Rio19971977NoCutaneous (also BM)7mNAPDKishimoto199712/FYesCutaneous (also BM)7mNAPDCastagnola199723/MNoCNS11mYesDeath (PD)Maloisel199734/MYesColonNAYesDeath (PD)Forrest199734/MYesTesticle120mYesDeath (PD)Liso199884/MNoCutaneous14mYesDeath (PD)Liso199822/FNoCutaneous14mYesDeath (PD)Liso199830/MYesCNS (CSF+)10mYesCR (12m)Liso199845/MYesCutaneous10mYesCR (20m)Liso199845/MYesCutaneous0mYesCR (31m)Present case37/FYesCNS (cSF+)1,5mYesDeath (PD)Present case45/MYesCutaneous31mYesDeath (PD)	Evans	1997	23/M	Yes	CNS	15m	Yes	Death (GVHD)
Evans     1997     49/F     Yes     CNS(CSF+)     NA     Yes     Death (PD)       Molero     1997     40/M     Yes     CNS(CSF+)     13m     Yes     Death (PD)       Ueda     1997     66/M     Yes     Cutaneous (also BM)     7m     Yes     NA       Del Rio     1997     28/F     No     Cutaneous (also BM)     7m     NA     PD       Kishimoto     1997     28/F     No     Cutaneous (also BM)     7m     NA     PD       Castagnola     1997     11/F     Yes     Cutaneous (also BM)     7m     NA     PD       Castagnola     1997     24/M     No     Cutaneous     2.3m     Yes     Death (PD)       Gastagnola     1997     34/M     Yes     Colon     NA     Yes     Death (PD)       Liso     1998     17/M     No     Lymph node     25m     Yes     Death (PD)       Liso     1998     8//M     No     Cutaneous     14m     Yes     Death (PD)	Evans	1997	22/M	Yes	Bone (mastoid)	6m	Yes	CR (NA)
Molero     1997     40/M     Yes     CNS(CSF+)     13m     Yes     Death (PD)       Ueda     1997     66/M     Yes     Cutaneous (also BM)     7m     Yes     NA       Del Rio     1997     28/F     No     Cutaneous (also BM)     7m     VA     PD       Kishimoto     1997     28/F     No     Cutaneous (also BM)     7m     NA     PD       Castagnola     1997     28/F     No     Cutaneous     2-3m     Yes     Death (PD)       Maloisel     1997     23/M     No     Colon     NA     Yes     Death (PD)       Maloisel     1997     34/M     Yes     Testicle     120m     Yes     Death (PD)       Liso     1998     17/M     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     48/M     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     25/F     No     Cutaneous     12m     Yes     Death (PD)	Evans	1997	49/F	Yes	CNS(CSF+)	NA	Yes	Death (PD)
Ueda     1997     66/M     Yes     Cutaneous (also BM)     7m     Yes     NA       Del Rio     1997     28/F     No     Cutaneous (also BM)     7m     NA     PD       Kishimoto     1997     28/F     No     Cutaneous (also BM)     7m     NA     PD       Castagnola     1997     11/F     Yes     Cutaneous     2-3m     Yes     PD       Castagnola     1997     23/M     No     CNS     11m     Yes     Death (PD)       Maloisel     1997     34/M     Yes     Colon     NA     Yes     Death (PD)       Liso     1998     34/M     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     48/M     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     22/F     No     Cutaneous     12m     Yes     Death (CPD)       Liso     1998     30/M     Yes     CNS (CSF+)     9m     Yes     CR (20m)	Molero	1997	40/M	Yes	CNS(CSF+)	13m	Yes	Death (PD)
Del Rio     1997     28/F     No     Cutaneous (also BM)     7m     NA     PD       Kishimoto     1997     11/F     Yes     Cutaneous     2-3m     Yes     PD       Castagnola     1997     12/M     No     CNS     11m     Yes     PD       Maloisel     1997     42/M     Yes     Colon     NA     Yes     Death(PD)       Forrest     1997     42/M     Yes     Colon     NA     Yes     Death(PD)       Liso     1998     17/M     No     Lymph node     25m     Yes     Death (PD)       Liso     1998     48/M     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     22/F     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     30/M     Yes     CNS (CSF+)     9m     Yes     CR (20m)       Liso     1998     4/F     Yes     Cutaneous     0m     Yes     CR (20m)       Liso	Ueda	1997	66/M	Yes	Cutaneous (also BM)	7m	Yes	NA
Kishimoto     1997     11/F     Yes     Cutaneous     2-3m     Yes     PD       Castagnola     1997     23/M     No     CNS     11m     Yes     Death (PD)       Maloisel     1997     23/M     No     CNS     11m     Yes     Death (PD)       Forrest     1997     24/M     Yes     Colon     NA     Yes     Death (PD)       Forrest     1997     34/M     Yes     Testicle     120m     Yes     Death (PD)       Liso     1998     84/M     No     Lymph node     25m     Yes     Death (PD)       Liso     1998     82/F     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     22/F     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     22/F     No     Cutaneous     12m     Yes     Death (PD)       Liso     1998     34/M     Yes     CNS (CSF+)     9m     Yes     CR (20m)       Lis	Del Río	1997	28/F	No	Cutaneous (also BM)	7m	NA	PD
Castagnola     1997     23/M     No     CNS     11m     Yes     Death (PD)       Maloisel     1997     42/M     Yes     Colon     NA     Yes     Death (PD)       Forrest     1997     34/M     Yes     Colon     NA     Yes     Death (PD)       Liso     1998     34/M     Yes     Testicle     120m     Yes     Death (PD)       Liso     1998     48/M     No     Lymph node     25m     Yes     Death (PD)       Liso     1998     82/F     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     22/F     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     30/M     Yes     Cutaneous     12m     Yes     Death (PD)       Liso     1998     30/M     Yes     CNS (CSF+)     9m     Yes     CR (20m)       Liso     1998     45/M     Yes     Cutaneous     0m     Yes     CR (20m)       Liso	Kishimoto	1997	11/F	Yes	Cutaneous	2-3m	Yes	PD
Maloisel     1997     42/M     Yes     Colon     NA     Yes     Death(PD)       Forrest     1997     34/M     Yes     Testicle     120m     Yes     Death(PD)       Liso     1998     17/M     No     Lymph node     25m     Yes     Death (PD)       Liso     1998     48/M     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     22/F     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     22/F     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     30/M     Yes     CNS (CSF+)     9m     Yes     CR (20m)       Liso     1998     45/M     Yes     Cutaneous     0m     Yes     CR (20m)       Liso     1998     45/M     Yes     Cutaneous     0m     Yes     CR (20m)       Liso     1998     45/M     Yes     Cutaneous     0m     Yes     CR (20m)       Present	Castagnola	1997	23/M	No	CNS	11m	Yes	Death (PD)
Forrest     1997     34/M     Yes     Testicle     120m     Yes     Death (PD)       Liso     1998     17/M     No     Lymph node     25m     Yes     Death (PD)       Liso     1998     48/M     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     22/F     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     25/F     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     32/M     Yes     Cutaneous     14m     Yes     Death (PD)       Liso     1998     30/M     Yes     CNS (CSF+)     9m     Yes     CR (20m)       Liso     1998     45/M     Yes     Cutaneous     10m     Yes     CR (20m)       Liso     1998     45/M     Yes     Cutaneous     0m     Yes     CR (31m)       Present case     37/F     Yes     CNS (cSF+)     1,5m     Yes     Death (PD)       Present case	Maloisel	1997	42/M	Yes	Colon	NA	Yes	Death(PD)
Liso     1998     17/M     No     Lymph node     25m     Yes     Death (PD)       Liso     1998     48/M     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     22/F     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     25/F     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     25/F     No     Cutaneous     12m     Yes     Death (PD)       Liso     1998     30/M     Yes     CNS (CSF+)     9m     Yes     CR (12m)       Liso     1998     44/F     Yes     Cutaneous     10m     Yes     CR (20m)       Liso     1998     44/F     Yes     Cutaneous     0m     Yes     CR (31m)       Present case     37/F     Yes     CNS (cSF+)     1,5m     Yes     Death (PD)       Present case     45/M     Yes     CNS (cSF+)     1,5m     Yes     Death (PD)	Forrest	1997	34/M	Yes	Testicle	120m	Yes	Death (PD)
Liso     1998     48/M     No     Cutaneous     14m     Yes     Death (PD)       Liso     1998     22/F     No     Cutaneous     14m     Yes     Death (VD)       Liso     1998     22/F     No     Cutaneous     12m     Yes     Death (VD)       Liso     1998     30/M     Yes     CNS (CSF+)     9m     Yes     CR (12m)       Liso     1998     44/F     Yes     Cutaneous     10m     Yes     CR (20m)       Liso     1998     44/F     Yes     Cutaneous     0m     Yes     CR (20m)       Liso     1998     45/M     Yes     Cutaneous     0m     Yes     CR (20m)       Present case     37/F     Yes     CNS (CSF+)     1,5m     Yes     Death (PD)       Present case     45/M     Yes     CNS (csF+)     1,5m     Yes     Death (PD)	Liso	1998	17/M	No	Lymph node	25m	Yes	Death (PD)
Liso     1998     22/F     No     Cutaneous     14m     Yes     Death (GVHD)       Liso     1998     25/F     No     Cutaneous     12m     Yes     Death (PD)       Liso     1998     30/M     Yes     CNS (CSF+)     9m     Yes     CR (20m)       Liso     1998     44/F     Yes     Cutaneous     10m     Yes     CR (20m)       Liso     1998     45/M     Yes     Cutaneous     0m     Yes     CR (20m)       Liso     1998     45/M     Yes     Cutaneous     0m     Yes     CR (31m)       Present case     37/F     Yes     CNS (CSF+)     1,5m     Yes     Death (PD)       Present case     45/M     Yes     CNS (extradural mass)     31m     Yes     Death (PD)	Liso	1998	48/M	No	Cutaneous	14m	Yes	Death (PD)
Liso     1998     25/F     No     Cutaneous     12m     Yes     Death (PD)       Liso     1998     30/M     Yes     CNS (CSF+)     9m     Yes     CR (12m)       Liso     1998     44/F     Yes     Cutaneous     10m     Yes     CR (20m)       Liso     1998     45/M     Yes     Cutaneous     0m     Yes     CR (31m)       Present case     37/F     Yes     CNS (CSF+)     1,5m     Yes     Death (PD)       Present case     45/M     Yes     CNS (extradural mass)     31m     Yes     Death (PD)	Liso	1998	22/F	No	Cutaneous	14m	Yes	Death (GVHD)
Liso     1998     30/M     Yes     CNS (CSF+)     9m     Yes     CR (12m)       Liso     1998     44/F     Yes     Cutaneous     10m     Yes     CR (20m)       Liso     1998     45/M     Yes     Cutaneous     0m     Yes     CR (31m)       Present case     37/F     Yes     CNS (CSF+)     1,5m     Yes     Death (PD)       Present case     45/M     Yes     CNS (extradural mass)     31m     Yes     Death (PD)	Liso	1998	25/F	No	Cutaneous	12m	Yes	Death (PD)
Liso     1998     44/F     Yes     Cutaneous     10m     Yes     CR (20m)       Liso     1998     45/M     Yes     Cutaneous     0m     Yes     CR (31m)       Present case     37/F     Yes     CNS (CSF+)     1,5m     Yes     Death (PD)       Present case     45/M     Yes     CNS (extradural mass)     31m     Yes     Death (PD)	Liso	1998	30/M	Yes	CNS (CSF+)	9m	Yes	CR (12m)
Liso     1998     45/M     Yes     Cutaneous     Om     Yes     CR (31m)       Present case     37/F     Yes     CNS (CSF+)     1,5m     Yes     Death (PD)       Present case     45/M     Yes     CNS (extradural mass)     31m     Yes     Death (PD)	Liso	1998	44/F	Yes	Cutaneous	10m	Yes	CR (20m)
Present case     37/F     Yes     CNS (CSF+)     1,5m     Yes     Death (PD)       Present case     45/M     Yes     CNS (extradural mass)     31m     Yes     Death (PD)	Liso	1998	45/M	Yes	Cutaneous	0m	Yes	CR (31m)
Present case 45/M Yes CNS (extradural mass) 31m Yes Death (PD)	Present case		37/F	Yes	CNS (CSF+)	1,5m	Yes	Death (PD)
	Present case		45/M	Yes	CNS (extradural mass)	31m	Yes	Death (PD)

A/S: age and sex; ATRA: prior therapy with ATRA; Interval: months from diagnosis to extramedullary disease; Cyto: t(15;17)/PML-RARα fusion protein. BM: bone marrow; CSF: cerebrospinal fluid; PD: progressive disease; NA: not available.

underwent an allogeneic bone marrow transplantation (BMT). Twenty months after the BMT he developed a spinal cord syndrome due to a paravertebral mass. Biopsy of the tumor revealed promyelocytes with a PML-RAR $\alpha$  rearrangement (bcr-1). Despite treatment with ATRA and local radiotherapy, the patient died from disease progression.

Extramedullary involvement, including CNS infiltration, has been rarely reported in APL. It usually occurs