

## Chloroma- Myelosarcoma or Leukemic Nerve Tumor?

Grisold W\*, Meng S and Grisold A

Department Neurology, KFJ Hospital and Medical University of Vienna, Austria

\*Corresponding author: Grisold W, Department Neurology, KFJ Hospital Vienna, Austria, Tel: +43(0)1601912050; E-mail: [grisoldw@gmail.com](mailto:grisoldw@gmail.com)

Received date: July 27, 2016; Accepted date: July 28, 2016; Published date: July 30, 2016

Copyright: © 2016 Grisold W et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

**Keywords:** Chloromas; Myelosarcoma; Leukemic tumors

### Introduction

Following our review on leukemia and the nervous system we would like to draw your attention to a rare manifestation of leukemia termed historically chloroma (CH) or myelosarcoma, which can rarely affect the cranial nerves and the peripheral nerves. These rare associations of leukemia and the peripheral nervous system, can occur in all stages of leukemia, as the first manifestation [1], as recurrence [2] and post bone marrow transplant. Most frequently CH are seen in AML, less frequent CML, but also in OMF. They can also appear isolated, and leave room for speculations why leukemia can convert from a liquid into a solid tumor, and why the presentation can be in peripheral nerves. Chloromas, myelosarcomas or leukemic tumors are rare [3-5] and present at many sites of the body. Compared to lymphomas focal nerve lesion in leukemia are rare [6].

For this review the presentation of CH in the peripheral nervous system, including the cranial nerves was chosen.

Peripheral nerve and CN tumors are rare. In leukemia the most frequent PNS lesions are meningeal involvement (LC), whereas neoplastic infiltration or compression of nerves by solid masses of leukemia is rare. Affection of CN and peripheral nerves is less frequently observed in leukemia, than in lymphoma.

### Leptomeningeal involvement or not?

Meningeal spread of leukemia is not infrequent. In particular the triad between CNS, CN and radicular symptoms is characteristic. CH do not necessarily involve the meninges, however proximity of CH to the meninges either intracranially or spinal does not exclude an additional LC. The following considerations focus on CH.

### Head and cranial nerves

CH can present in the orbit [7,8] infiltrate the optic nerve [9] affect the cavernous sinus [10] and causing focal CN damage, and also cause isolated hearing loss [10]. In addition mastoid or temporal bone lesions can cause CN dysfunction. CN can also be involved in leukemia in meningeal seeding (LC), dural involvement and also solid leukemic tumors presenting orbit, the optic nerve, in the cavernous sinus, with hearing loss, have been reported.

### Roots

The nerve roots can be the site of leukemic infiltration also presenting as a mass lesion. The involvement of the cauda equina by a leukemic mass has been observed [11-13].

### Plexus

The brachial plexus as well as the lumbar and sacral plexus can be involved by CH. Due to modern imaging techniques the detection rate has improved. As the brachial, lumbar and sacral plexus can be easily examined. From the literature, the brachial plexus [14-16] seems more involved than the lumbar and sacral plexus. However this may also be an artifact due more specific symptom based evaluation of the brachial plexus. The lumbar plexus can present with low back pain [17]. Also the sacrum can be the site CH with local nerve lesions [18-20].

### Mononeuropathies

The presentation of CH in peripheral nerves presenting as mononeuropathies is rare. In some conditions also the term neuroleukemiosis has been suggested [21]. Leukemia can infiltrate nerves or present as a diffuse parenchymatous tumor also affecting nerves [22] or presenting as a solid nerve tumor.

On the UE both the median and the ulnar nerve have been described [21] and ulnar nerve [23]. In the lower extremities most cases have been observed in the femoral [24] and sciatic [25-27] and peroneal [28] nerve.

In the clinical setting several differential diagnoses ranging from entrapment neuropathies, rare isolated effects of chemotherapy and peripheral nerve tumors, as amyloidomas and rare leukemic deposition have to be considered. This is in particular difficult in cases in remission or after transplantation. In leukemia also coagulopathies can cause focal hemorrhages into peripheral nerves resulting in painful mononeuropathies.

### Muscle

Also skeletal muscle can be the site of CHs [29], Muscle: masseteric muscle [30,31].

### Discussion

The occurrence of a solid mass as presentation of leukemia (CH), is a rare event which can occur as a presentation, during the course of leukemia, as a relapse or as a complication of bone marrow transplant.

Imaging with MR, US and PET [16,32] has facilitated the detection, which may point of a more frequent appearance (However precise clinical criteria for the appearance of CH in ultrasound and MR are lacking).

The appearance of leukemia and CH in peripheral nerves is interesting. It has been proposed that the peripheral nerves and the peripheral nerve blood barrier may be an ideal situation to leukemic cells to survive in a sanctuary. However in most cases of leukemia the CN and peripheral nerves are spared from tumor infiltration, except

the in intra - meningeal part, where LC is a frequent event. Another interesting aspect is the transformation of a liquid cancer into a solid cancer, at times isolated as a solid cancer, without the liquid presentation.

The appearance of CH is often uncharacteristic and may require biopsy. Peripheral tumors are rare the differential diagnostic approach needs to include several other differential diagnoses in particular if the CH is the first manifestation of leukemia. Once leukemia has been diagnosed this is more likely in acute and myeloid leukemia, and it has been described as recurrence or after BMT.

## References

1. Hurwitz BS, Sutherland JC, Walker MD (1970) Central nervous system chloromas preceding acute leukemia by one year. *Neurology* 20: 771-775.
2. Zheng C, XL, Zhu W, Cai X, Wu J, et al. (2014) Tailored central nervous system- directed treatment strategy for isolated CNS recurrence of adult acute myeloid leukemia. *Hematology* 19: 208-212.
3. Pochedly C (1975) Neurologic manifestations in acute leukemia. III. Peripheral neuropathy and chloroma. *N Y State J Med* 75: 878-882.
4. Imrie KR, Kovacs MJ, Selby D, Lipton, Patterson BJ, et al. (1995) Isolated chloroma: the effect of early antileukemic therapy. *Ann Intern Med* 123: 351-353.
5. Paydas S, Zorludemir S, Ergin M (2006) Granulocytic sarcoma: 32 cases and review of the literature. *Leuk Lymphoma* 47: 2527-2541.
6. Grisold W, Briani C, Vass A (2013) Malignant cell infiltration in the peripheral nervous system. *Handb Clin Neurol* 115: 685-712.
7. Kumar J, Seith A, Bakhshi S, Kumar R, Kumar A, et al. (2007) Isolated granulocytic sarcoma of the orbit. *Eur J Haematol* 78: 456.
8. Cavdar AO, Arcasoy A, Babacan E, Gozdasoglu S, Topuz U, et al. (1978) Ocular granulocytic sarcoma (chloroma) with acute myelomonocytic leukemia in Turkish children. *Cancer* 41: 1606-1609.
9. Shah P, Yohendran J, Lowe D, McCluskey P (2012) Devastating bilateral optic nerve leukaemic infiltration. *Clin Experiment Ophthalmol* 40: e114-115.
10. Alami BMM (2014) Acute lymphoblastic leukemia revealed by an invasion of the cavernous sinus in a young man. *Pan Afr Med J* 98.
11. Gokcan MK, Batikhan H, Calguner M, Tataragasi AI (2006) Unilateral hearing loss as a presenting manifestation of granulocytic sarcoma (chloroma). *Otol Neurotol* 27: 106-109.
12. Smith TR, Slimack N, McClendon J, Wong A, Fessler RG, et al. (2012) Low back pain and lumbar radiculopathy as harbingers of acute myeloid leukemia recurrence in a patient with myeloid sarcoma. *J Clin Neurosci* 19: 1040-1041.
13. Buakhao JTA (2011) Cauda equina involvement in acute myeloid leukemia relapse. *J Med Assoc Thai* 94: 1271-1275.
14. Onal IK, Göker H, Büyüksayık Y, Ozçakar L (2006) Cauda equina syndrome as a rare manifestation of leukemia relapse during post- allograft period. *J Natl Med Assoc* 98: 808-810.
15. Ha Y, Sung DH, Yoonhong MD, Park MD, Du HK, et al. (2013) Brachial Plexopathy due to Myeloid Sarcoma in a Patient With Acute Myeloid Leukemia After Allogenic Peripheral Blood Stem Cell Transplantation. *Ann Rehabil Med* 37: 280-285.
16. Heckl S, Horger M, Faul C, Ebrahimi A, Ioanovicu SD, et al. (2014) Myeloid sarcoma of nervous plexus - infiltration of the nerve plexus by extramedullary manifestation of acute myeloid leukemia. *Rofa* 186: 1059-1062.
17. Mauermann ML, Angius D, Spinner RJ, Letendre LJ, Amrami KK, et al. (2008) Isolated granulocytic sarcoma presenting as a brachial plexopathy. *J Peripher Nerv Syst* 13: 153-156.
18. Boleto G, Michel M, Salam N, Eschard JP, Salmon JH, et al. (2016) Low back pain and femoral neuralgia revealing myeloid sarcoma with megakaryocytic differentiation. *Joint Bone Spine*.
19. Novick SL, Nicol TL, Fishman EK (1998) Granulocytic sarcoma (chloroma) of the sacrum: initial manifestation of leukemia. *Skeletal Radiol* 27: 112-114.
20. Massoud M, Del Bufalo F, Musolino AMC, Schingo PM, Gaspari S, et al. (2016) Myeloid Sarcoma Presenting as Low Back Pain in the Pediatric Emergency Department. *J Emerg Med*.
21. Stork JT, Cigtay OS, Schellinger D, Jacobson RJ (1984) Recurrent chloromas in acute myelogenous leukemia. *AJR Am J Roentgenol* 142: 777-778.
22. Wang T, Miao Y, Meng Y, Li A (2015) Isolated leukemic infiltration of peripheral nervous system. *Muscle Nerve* 51: 290-293.
23. Warme B, Sullivan J, Tigrani DY, Fred DM (2009) Chloroma of the forearm: a case report of leukemia recurrence presenting with compression neuropathy and tenosynovitis. *Iowa Orthop J* 29: 114-116.
24. Bakst R, Jakubowski A, Yahalom J (2011) Recurrent neurotropic chloroma: report of a case and review of the literature. *Adv Hematol*.
25. Bakst R, Wolden S, Yahalom J (2012) Radiation therapy for chloroma (granulocytic sarcoma). *Int J Radiat Oncol Biol Phys* 82: 1816-1822.
26. Stillman MJ, Christensen W, Payne R, Foley KM (1988) Leukemic relapse presenting as sciatic nerve involvement by chloroma (granulocytic sarcoma). *Cancer*. 62: 2047-2050.
27. Mosch A, Kazzaz BA (1991) Intradural granulocytic sarcoma: a rare cause of sciatic pain. *Clin Neurol Neurosurg* 93: 341-344.
28. Eusebi V, Bondi A, Cancellieri A, Canedi L, Frizzera G, et al. (1990) Primary malignant lymphoma of sciatic nerve. Report of a case. *Am J Surg Pathol* 14: 881-885.
29. Aregawi, Sherman JH, Douvas, Burns TM, Schiff D (2008) Neuroleukemiosis: case report of leukemic nerve infiltration in acute lymphoblastic leukemia. *Muscle Nerve* 38: 1196-1200.
30. Song-Mee Cho, WH J (2009) Granulocytic Sarcoma in the Leg Mimicking Hemorrhagic Abscess. *JKSMRM* 13: 88-92.
31. Bassichis B, McClay J, Wiatrak B (2000) Chloroma of the masseteric muscle. *Int J Pediatr Otorhinolaryngol* 53: 57-61.
32. Chhabra A, Thakkar RS, Andreisek G, Chalian M, Belzberg AJ, et al. (2013) Anatomic MR imaging and functional diffusion tensor imaging of peripheral nerve tumors and tumorlike conditions. *AJNR Am J Neuroradiol* 34: 802-807.