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Chloroma- Myelosarcoma or Leukemic Nerve Tumor?

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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 covert 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 the site of leukemic infiltration also presenting as a mass lesion. The involvement of the cauda equine 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 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: massteric 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.

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