Intracranial Chloroma (Granulocytic Sarcoma) by Lymphocytic Leukemia

Ho Seak Jeong, M.D., Moo Seong Kim, M.D., Yong Tae Jung, M.D., Jae Hong Sim, M.D.
Department of Neurosurgery, Inje University Busan Paik Hospital, Busan, Korea

Intracranial chloroma may occur in leukemia, although they are rare. A 23-year-old female complained diplopia. Brain magnetic resonance MR imaging showed tumors in the both cavernous sinus, both tentorial and anterior falx. Gamma-Knife radiosurgery was performed with maximal dose; 200y, marginal dose; 10Gy. Peripheral blood smear revealed leukemia, and bone marrow aspiration biopsy showed acute lymphocytic leukemia. Two weeks later, MR image for the stereotactic biopsy noticed markedly decreased tumor size. Biopsy result was lymphocytic leukemia. She received conventional radiation therapy, chemotherapy, and bone marrow transplantation. Brain involvement by acute lymphocytic leukemia is very rare. Even though chloroma are sensitive to radiation therapy, prognosis is poor because of the gravity of the underlying disease and association with impending blast transformation. The authors reports a intracranial chloroma by acute lymphocytic leukemia.

KEY WORDS: Leukemia · Chloroma · Brain.

Introduction

Granulocytic sarcoma (chloroma) is a localized tumor mass composed of immature cells of the granulocytic series. Neiman, et al reported that the lesion was first described by Burns in 1811. The term "Chloroma" was given the lesion because such tumors often display a greenish color that fades on exposure to air. This color is due to the presence of myeloperoxidase (verdoperoxidase) in the tumor cells "Granulocytic sarcoma," the preferred term, was first used by Rappaport.

Since the association of granulocytic sarcoma with acute leukemia have been reported in patients with acute myeloid leukemia(AML), most notably in childhood, and frequently are noted only at autopsy. Central nervous system involvement may be either meningeal or parenchymal, although the overwhelming majority of cases are meningeal. Infiltration of the meninges by leukemic cells may affect the dura, the leptomeninges, or both, and may be diffuse or focal. In rare cases, leptomeningeal tumor may be focal instead of diffuse, and differentiation from parenchymal lesions may be difficult. Intracranial masses called chloroma or granulocytic sarcoma may occur in leukemia, although they are rare.

We experienced one patient affected by acute lymphoblastic leukemia(ALL) with granulocytic sarcoma (chloroma). This is a very rare chloroma case caused by ALL. We review radiological, pathological findings in chloroma, and followup result.

Case Report

A 23-year-old female patient was referred for neurosurgical evaluation due to diplopia. Brain magnetic resonance (MR) imaging showed multiple intracranial tumors in both cavernous sinus, both tentorial and anterior falx. Initial, she refused biopsy because she was young. She had received Gamma-Knife radiosurgery for tumors, with maximal dose; 20Gy, marginal dose; 10Gy. Stereotactic biopsy was done after Gamma-Knife radiosurgery. Stereotactic biopsy was underwent on left tentorium, three pieces of the specimen was obtained. The biopsy result was lymphocytic leukemia. The peripheral smear showed leukemia. After 1 month later, MRI showed marked decrease in tumor.
size. She had received conventional radiation therapy, 3 cycles of the chemotherapy regimen: cyclophosphamide 1,400mg, 6-mercaptopurine 90mg, cyclosporine 40mg, vincristine 2mg, L-asparaginase 9000U 6-mercaptopurine 90mg, intrathecal methotrexate 15mg was given, 1 month interval. 4 months later, she received bone marrow transplantation. There was no side effect. One and half year later now, she is well, and there has been no development of tumor recur.

Discussion

Chloromas (granulocytic sarcoma) are masses composed of immature granulocytic cells. The term 'chloroma' has been derived from the Greek word chloros (green) describing the lesion's typical greenish appearance. The color is due to high expression of myeloperoxidase, and is not always detectable because it rapidly fades after exposure to oxygen

Granulocytic sarcoma occurs primarily in patients with AML but may also arise in patients with myeloproliferative and myelodysplastic syndrome. Extracranial sites such as skin, bone, and soft tissue are far more commonly affected than the CNS. Chloromas were found in 3.1–9.1% of patients ultimately proven to have acute myelogenous leukemia (AML) or chronic myelogenous leukemia (CML).

Present signs and symptoms of cranial and intracranial granulocytic sarcomas are nonspecific. Cerebral angiography may show a focal mass with a meningial vascular supply, a dense vascular blush, and early venous filling. Precontrast computed tomography (CT) studies show a hyperdense or isodense focal mass with surrounding edema, and postcontrast CT demonstrates well-circumscribed, homogeneous tumor enhancement with or without a relatively hypodense center. Most of the lesions will have low signal intensity on the relatively T1-weighted images and high signal intensity on T2-weighted images.

On gross pathologic examination, the chloromas are usually firm, but not strong hard, invade into the surrounding tissue, and appear bright green to pale green to dull green in color. Chloromas arise most frequently in children and young adults.

The peak incidence is between 4 and 5 years of age; 60.1%
of cases are observed in patients less than 15 years of age.12, 9). In our case, patient’s age was 22 years.

In some cases, removal of tumor discovered early may lead
to a long symptom free period, and tumor response to ex-
ternal radiation has been well documented. Usually, the
disease is considered invariably fatal.2, 3). With a median survival
of 22 months, they are associated with a poor prognosis.2, 3)

Granulocytic sarcoma may be the first sign of relapse after
bone marrow transplantation or may precede the onset of
systemic disease.5, 11) Imrie et al.3) demonstrated that surgical
resection and/or irradiation, although highly effective in
local control, do not influence survival. The best results
were achieved with antileukemic chemotherapy containing
Ara-C in higher doses.5, 6) An equally favorable outcome of
the few reported patients with autologous and allogeneic
bone marrow transplantation after induction therapy suggests
an intensified consolidation to be useful.5, 11)

In our case, she received radiotherapy, GammaKnife ra-
diosurgery, chemotherapy, bone marrow transplantation.
Favorable result was obtained because she was young.
Chemotherapy, radiotherapy and bone marrow transplantation
affected her good result. In myeloblastic leukemia, CNS
involvement is often development, but in lymphoblastic leu-
kemia, CNS involvement is very rare with literature review.41)

Conclusion

Granulocytic sarcoma by acute lymphocytic leukemia is
very rare. Though very sensitive to radiation therapy,
prognosis is poor because the gravity of the underlying disease
and association with impending blast transformation. Despite
reports of complete dissolution of the intracranial tumors
following radiation therapy, patients tend to die, usually in
blast crisis, within months following discovery of the lesion
onset of neurologic symptoms. Early detection of the chloroma
may help the final outcome of the disease.

References
2. Bekassy AN, Hermans J, Gorin NC, Grauwohl A: Granulocytic sarcoma
after allogeneic bone marrow transplantation: a retrospective European
multicenter survey. Bone Marrow Transplant 17 : 801-808, 1996
4. Cavdar AO, Arcasoy O, Babacan E, Guzglasoglu S, Topuz U, Furaumu
JF: Ocular granulocytic sarcoma (chloroma) with acute myelomonocy-
5. Huter O, Brezinka C, Nadzhaur D, Schwaighofer H, Lang A, Nied-
erwieser D: Successful treatment of primary extramedullary leukemia of
the uterus with radical therapy, chemotherapy, autologous bone
marrow transplantation and prophylactic local irradiation. Bone Marrow
Transplant 18 : 663-664, 1996
sarcoma of the thoracic spine. J Korean Neurosurg Soc 22 : 853-858,
1993
8. Kao SCS, Yuh TC, Sato Y, Barloon TJ: Intracranial granulocytic sar-
coma (chloroma): M.R. findings. J Comput Assist Tomogr 11 : 938-941,
1987
study of granulocytic sarcoma (chloroma) in patients with myelogenous
1973
10. Must HB, Moloney WC: Chloroma and other myeloblastic tumors.
Blood 42 : 721-728, 1973
Granulocytic sarcoma: a clinicopathologic study of 61 biopsied cases.
12. Pomerantz SJ, Hawkins HH, Towbin R, Lisberg WN, Clark RA:
Granulocytic sarcoma (chloroma): CT manifestations. Radiology
155 : 167-170, 1985
CML presenting with hypercalcemia and a mediastinal mass. Acta
14. Rappaport H: Tumors of the hematopoietic system, atlas of tumor
pathology, section III, Fascicle 8. Armed Forces Institute of Pathology,
15. Sandhu GS, Gonzalez-Garcia J, Elexoru-Caminya JA: Granulocytic sarcoma
presenting as cauda equina syndrome. Clin Neurol Neurosurg
100 : 205-208, 1998
16. Sowers JJ, Moody DM, Naidich TP, Ball MR, Laster DW, Leeds NE:
Radiographic features of granulocytic sarcoma (chloroma). J Comput
Assist Tomogr 3 : 226-233, 1979