CASE REPORT

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Intracranial Chloroma(Granulocytic Sarcoma) by Lymphocytic Leukemia

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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; 20Gy, 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[11].

The term "Chloroma" was given the lesion because such tumors often display a greenish color that fades on exposure to air[11]. This color is due to the presence of myeloperoxidase (verdoperoxidase) in the tumor cells "Granulocytic sarcoma," the preferrred term, was first used by Rappaport[14].

Since the association of granulocytic sarcoma with acute leukemia have been reported in patients with acute myeloid leukemia(AML), most notably in childhood[4], and frequently are noted only at autopsy[9].

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[9].

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 cavenous 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 1month later, MRI showed marked decrease in tumor

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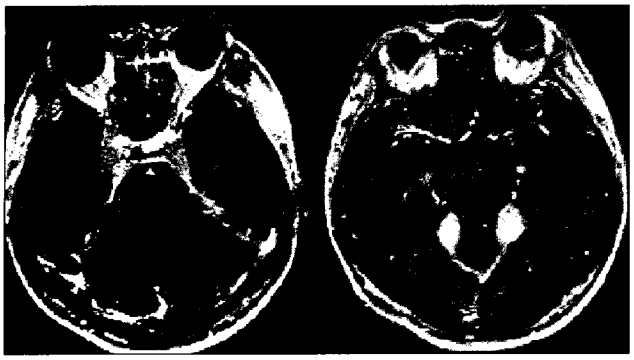

Fig. 1. Initial enhanced T1-weighted magnetic resonance images showing the both cavernous sinus, both tentorial and anterior falx tumors.

size. She had received conventional radiation therapy. 3cycles of the chemotherapy regimen:cyclophosphamide 1,400mg, 6-mercaptopurine 90mg, cytarabine 110mg, vincristine 2mg, L-asparaginase 9000U 6-mercaptopurine 90mg, intrathecal methotrexate 15mg was given, 1month interval. 4months 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 facets after exposure to oxygen[15].

Granulocytic sarcoma occurs primarily in patients with AML but may also arise in patients with myeloproliferative and myelodysplastic syndrome[11,13]. Extracranial sites such as skin, bone, and soft tissue are far more commonly affected than the CNS[11,13]. Chloromas were found in 3.1~9.1% of patients ultimately proven to have acute myelogenous leukemia(AML) or chronic myelogenous leukemia(CML)[2,3,9,10,16].

Present signs and symptoms of cranial and intracranial granulocytic sarcomas are nonspecific. Cerebral angiogrophy may show a focal mass with a meningial vascular supply, a dense vascular blush, and early venous filling[16]. Precontrast computed tomography(CT) studies show a hyperdense or isodense focal mass with variable surrounding edema, and postcontrast CT demonstrates well-circumscribed, homogenous tumor enhancement with or without a relatively hypodense center[16]. Most of the lesions will have low signal intensity on the relatively T1-weighted images and high signal intensity on T2-weighted images[16].

On gross pathologic examination, the chloromas are usually firm, but not strong hard, invade into the surrounding tissue,

Fig. 2. Stereotactic biopsy show : Portion of wall formed by fibrous tissue, containing a dense round cell infiltrate and lined by stratified squamous, nonkeratinized epithelium(H&E, original magnification ×40). Hematoxylin and eosin stain of the resected tumor reveals the presence of abundant histiocytic-like cells, with prominent eosinophilia and nuclear pleomorphism. The pathologic specimens were ultimately interpreted as granulocytic sarcoma(×400).

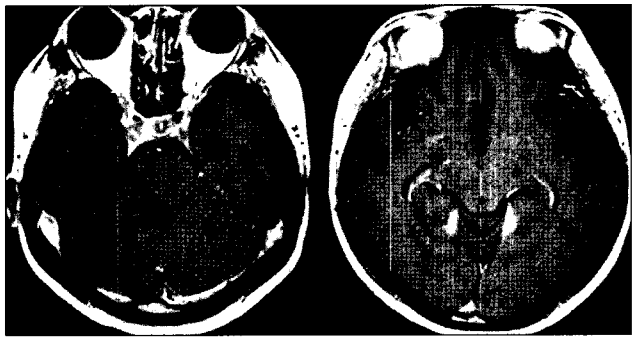

Fig. 3. One month later, enhanced T1-weighted magnetic resonance images after Gamma-Knife radiosurgery showing markedly decreased tumors size.

and appear bright green to pale green to dull green in color[15]. Chloromas arise most frequently in children and young adults[9].

The peak incidence is between 4 and 5years of age; 60.1%

of cases are observed in patients less than 15years of age[1,7-9]. In our case, patient's age was 22years.

In some cases, removal of tumor discovered early may lead to a long symptom free period, and tumor response to external radiation has been well documented. Usually, the disease is considered invariably fatal[2,3]. With a median survial of 22months, 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[1,11]. Imrie et al.[6] demonstrated that surgical resection and/or irradiation, although highly effective in local control, do not influence survival. The best results were achived with antileukemic chemotherapy containing Ara-C in higher doses[5,6]. An equally favorable outcome of the few reported patients with autologous and allogeneous bone marrow transplantation after induction therapy suggests an intensified consolidation to be useful[5,11].

In our case, she received radiotherapy, GammaKnife radiosurgery, chemotherapy, bone marrow transplantation. Favorable result was obtained because she was young. Chemotherapy, radiotherapy and bone marrow transplantation influenced her good result. In myeloblastic leukemia, CNS involvement is often development, but in lymphoblastic leukemia, CNS involvement is vere rare with literature review[11].

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.

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