The current role of radiotherapy in chloroma: Report of two cases and review of the literature.

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ABSTRACT: Chloroma (myeloid sarcoma) is a rare extramedullary manifestation of haematologic malignancy, most commonly acute myeloid leukemia(AML).It can occur in association with Myelogenous leukemia myeloproliferative disorders, and myelodysplasia.Myeloid sarcoma has different modalities of presentation and can affect any organ. We report two cases which has been irradiated in the Radiotherapy Oncology Department in Aristotle University Hospital AHEPA of Thessaloniki.

Key Words: Chloroma, Myeloid sarcoma, Acute myeloid leukemia, Radiotherapy.

INTRODUCTION

Chloroma (also known as granulocytic sarcoma or myeloid sarcoma) is a rare, extramedullary tumor composed of immature granulocytic cells. It was first described in 1811 and coined "chloroma" by King in 1853 because of its green color. Its relationship with Leukemia was later established in 1893[3].

Chloromas are reported in 2,5% - 9,1%[1,2,3] of patients with acute myeloid leukemia(AML) and occur concomitamtly, following, or rarely antedating the onset of leukemia[4]. The presence of an extramedullary relapse of leukemia is often associated with a poor prognosis[5]. The most common locations of chloromas are skin, soft tissue, bone, periosteum, and Lymph nodes[5]. Central nervous system (CNS) involvement is rare but has been described in numerous case reports[6,7,8]. Myeloid sarcoma can also develop in leukemia patients concurrently with or after diagnosis,or as manifestation of disease relapse[15,16]. It is rare for the clinical manifestation of a myeloid sarcoma to include signs of nerve root entrapment. In the current cases we present two patients with chloroma who had painful mass and edema in soft tissue and lymph nodes.

CASE REPORT

Two patients with chloroma came to our Radiotherapy Oncology Department for irradiation. The first patient was a 49 year old woman who developed pain and edema in her right low foot, 8 months ago. CT (Computer Tomography)revealed involvement by a presumed chloroma, mass in madibulum, left sacrum, O5 vertebra and right inguinale nodes (Figure 1). Biopsy of the mandibulum indicated myeloid sarcom mixed immunophenotyp B/myelogenen type. The patient was irradiated palliatively in her right inguinal nodes, with a total dose 2000cGy, in 5Fr with 400cGy/Fr (Figure 2). At the end of therapy she had decreased pain and reduction of edema in her right low foot. The CT after two months of irradiation showed reduction of mass (presumed chloroma) in her right inguinal nodes (Figure 3). There was no recurrence and the patient was with improvement and satisfying per-

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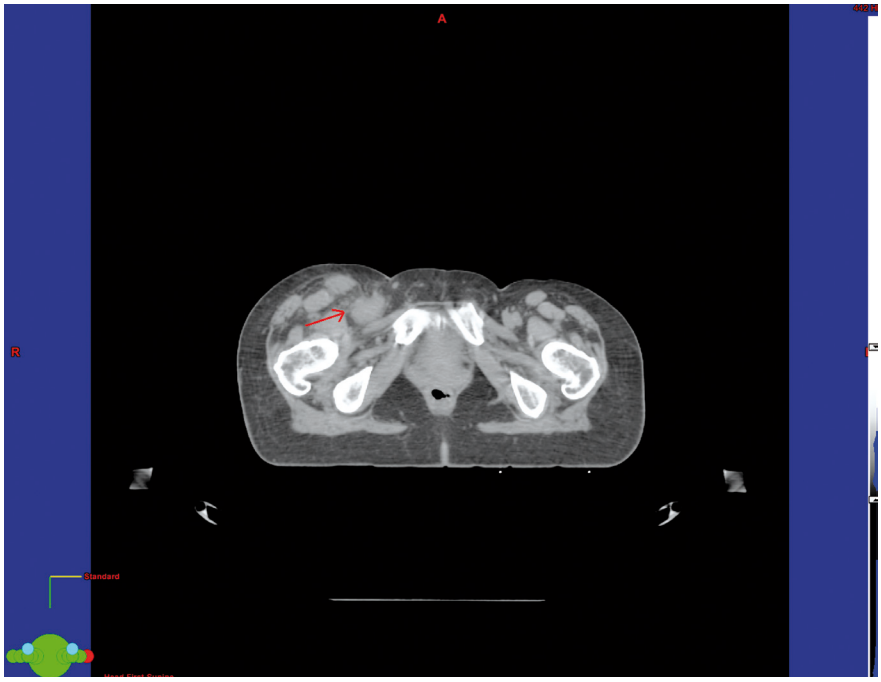

Figure 1. Involved region(inguinale nodes).

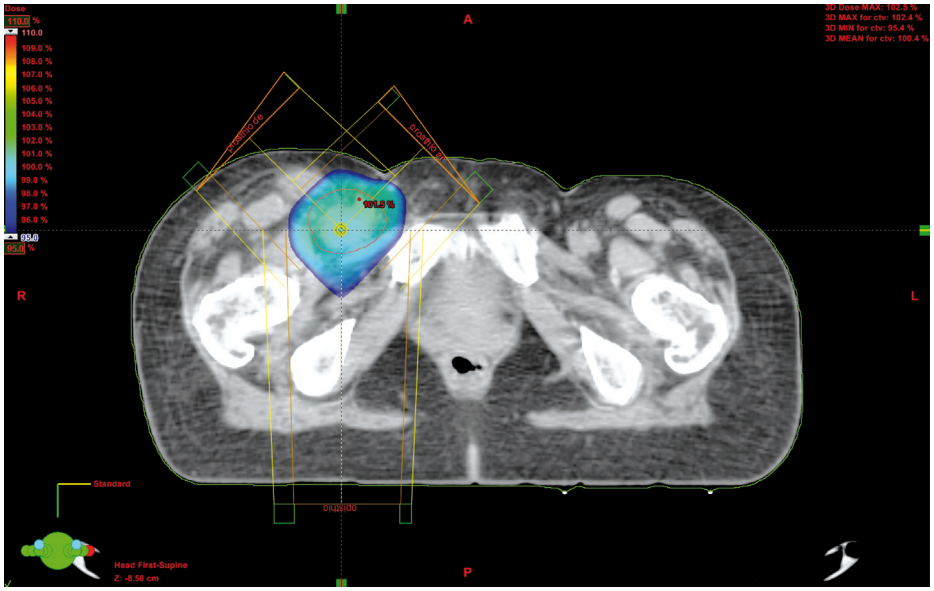

Figure 2. Fields for irradiation(right inguinale nodes).

formance status. The second patient, was an 70 year old woman, came in our radiotherapy department 6 months ago, with mass on the right neck and supraclavicular region. The biopsy proved also myeloid sarcoma. The patient developed pain and edema in these regions.She was irradiated palliatively in the neck and the supraclavicular region with dose 2000cGy, in 5Fr with 400Gy/Fr. At the end of therapy her mass had a significant remission during palpation. She was decreased pain and much better improvement. Both of them received radiotherapy with three fields, in the region of the chloromas and the radiotherapy was the treatment planning combined with chemotherapy in hematological department.

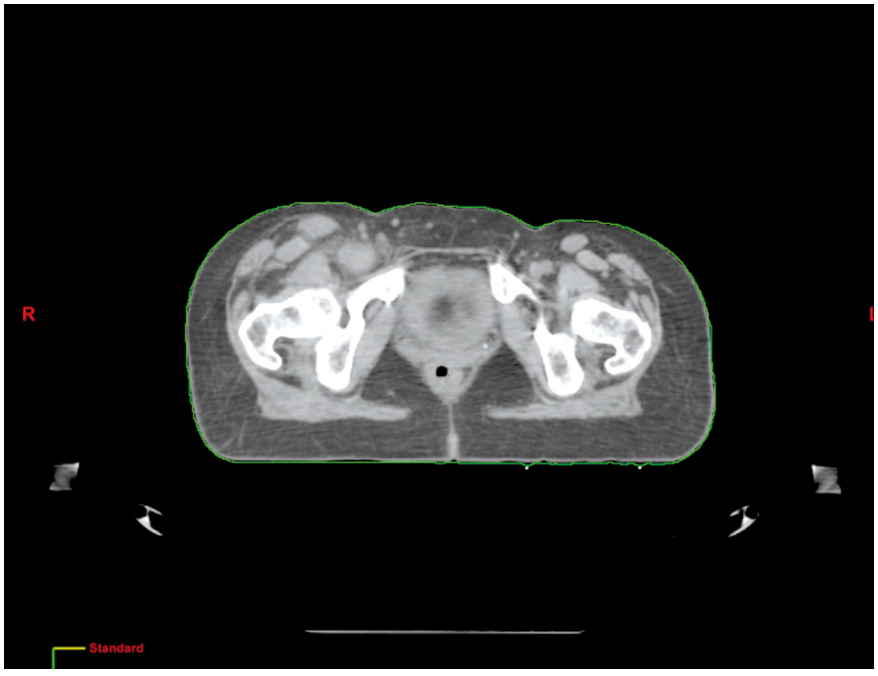

Figure 3. Reduction of chloroma after irradiation.

DISCUSSION

Chloromas' rare incidence, their often misdiagnosis[9] , and variable locations have resulted in limited clinical experience, and hence in a lack of consensus treatment guidelines. Myeloid sarcoma (chloroma) is a rare, malignant,and extramedullary solid tumour primarily associated with AML (Acute Myelogenous Leukemia), especially the M2 subtype, and it is often a precursor to the diagnosis of myelogenous leukemia or a manifestation of leukemia recurrence. Chloroma can also be found in multiple other myelodysplastic syndromes and can represent leukemic transformation[2]. Of nonleukemic patients who present with myeloid sarcoma, most will develop AML within 1 year (median 5 months)[11]. Chloroma has been reported in ≤9% of patients with AML,most commonly in the bone,periosteum, lymph nodes,skin,and multiple soft-tissue sites[2,12]. The tumour precedes systemic leukemia in 0,6% of cases [4]. Myeloid sarcoma is seen more often in men than in women as is generally the case in leukemia[13]. The tumour may appear at any age or localization. Because of the rarity of this disorder,large series of patients are seldom reported,and the prognosis and optimal treatment of patients presenting with MS are not clear.In general,the prognosis of myeloid sarcoma is usually considered to be poor[5]. Overall,the median survival is 7-20 months after the diagnosis of chloroma[11,14]. Considering the diagnosis of myeloid sarcoma at an early stage is important because of the therapeutic options in this potentially devastating disease. The role of radiation in their management has typically been palliative as low doses have resulted in excellent disease control and symptom relief. In contrast to older studies that support the use of at least 3000cGy, our experience with these cases and others suggests that 2000cGy-2400cGy is adequate[10] .

CONCLUSION

In conclusion,we believe that the value of radiation therapy was investigated in these patients with chloromas' treated at the Radiotherapy Department of Aristotelian University AHEPA Hospital. Such isolated recurrences may benefit from radiation therapy (low doses). This challenges the belief that chloromas always precede systemic relapse and suggests an evolving role for radiation therapy in the management of chloromas beyond palliation. Further,more comprehensive studies of chloromas are warranted to determine optimal management.

Ο θεραπευτικός ρόλος της ακτινοθεραπείας στο χλώρωμα: Αναφορά δυο περιπτώσεων και ανασκόπηση της βιβλιογραφίας.

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ΠΕΡΙΛΗΨΗ: Το χλώρωμα (μυελοειδές σάρκωμα) είναι μια σπάνια εξωμυελική εκδήλωση αιματολογικής κακοήθειας περισσότερο συχνή στην οξεία μυελογενή λευχαιμία (ΟΜΛ). Μπορεί να εμφανιστεί σε σχέση με τη μυελογενή λευχαιμία, μυελουπερπλαστικά σύνδρομα και διαταραχές, και μυελοδυσπλασία. Το μυελογενές σάρκωμα έχει διάφορες μορφές εκδήλωσης και μπορεί να προσβάλλει κάθε όργανο στο ανθρώπινο σώμα.

Λέξεις Κλειδιά: Χλώρωμα, Μυελοειδές σάρκωμα, Οξεία μυελογενής λευχαιμία, Ακτινοθεραπεία.

REFERENCES

1. P.I. Liu, T. Ishimaru, D.H McGregor, H.Okada, and A. Steer. "Autopsy study of granulocytic sarcoma (chloroma) in patients with myelogenous leukemia, Hiroshima-Nagasaki 1949-1969'', Cancer 1973; 31 vol, no4: 948-955.
2. R.S. Neiman, M. Barcos and C. Berard, ''Granulocytic sarcoma: a clinicopathologic study of 61 biopsied cases'', Cancer 1981; vol: 48, no6: 1426-1437.
3. P.H Wiernic and A. A. Serpick, "Granylocytic sarcoma (chloroma), "Blood, 1970: vol 35, no.3:361-369.
4. J.R. Krause, "Granulocytic sarcoma preceding acute leukemia. A report of six cases'', Cancer, 1979; vol 44, no3: 1017-1021.
5. S. Paydas, S. Zorludemir, and M. Ergin, "Granulocytic sarcoma: 32 cases and review of the literature'', Leukemia and Lymphoma 2006; vol. 47, no.12:2527:2541.
6. W.C. Verra, T.J. Snijders, T. Seute, K.S. Han, H.K. Nieuwenhuis, and G.J.Rutten, "Myeloid sarcoma presenting as a recurrent, multifocal nerve root entrapment syndrome'', Journal of Neuro-Oncology, 2009; vol. 91, no.1:59-62.
7. G.Widhalm, W. Dietrich, L. Mullauer, "Myeloid sarcoma with multiple lesions of the central nervous system in a patient without leukemia: case report'', Journal of Neurosurgery 2006; vol. 105, no.6:916-919.
8. W. Struhal, S. Oberndorfer, H. Lahrmann, "Myeloid sarcoma in the central nervous system: case report and review of the literature", Acta Clinica Croatica, 2008; vol. 47, no.1:19-24.
9. J.C. Byrd, W.J. Edenfield, D.J. Shields, and N.A. Dawson, "Extramedullary myeloid cell tumors in acute nonlymphocytic leukemia: a clinical review'', Journal of clinical Oncology, 1995; vol. 13, no.7:1800-1816.
10. L.Y. Chak, M.O. Sapozink, and R.S. Cox, "Extramedullary lesions in non-lymphocytic leukemia: results of radiation therapy'', International Journal of Radiation Oncology Biology Physics 1983; vol.9, no.8:1173-1176.
11. Breccia M, Mandelli F, Petti MC, et al Clinicopathological characteristics of myeloid sarcoma at diagnosis and during Follow-up: Report of 12 cases from a single institution. Leuk. Res. 2004; 28:1165-1169.
12. Meis JM, Butler JJ, Osborne BM, et al. Granulocytic sarcoma in nonleukemic patients. Cancer. 1986; 58: 2647-2709.
13. Mostafavi H, Lennarson PJ, Traynellis VC (2000) Granulocytic sarcoma of the spine. Neurosurgery 46:78-83.
14. Tsimberidou AM, Kantarjia HM, Estey E, et al. Outcome in patients with nonleukemic granulocytic sarcoma treated with chemotherapy with or without radiotherapy. Leukemia. 2003; 17:1100-1103.
15. P.H. Wiernik, R. De Bellis, P.Muxi, and J.P. Dutcher, "Extramedullary acute promyelocytic leukemia'', Cancer 1996, vol. 78, p.25.
16. M. Benekli, M.C. Savas, I.C. Haznedaroglou, and S.V. Dundar, "Granulocytic sarcoma in acute promyelocytic leukemia'', Leukemia and Lymphoma, 1996. vol. 22, no1-2, pp 183-185.