Intracranial chloroma in known case of Acute myeloblastic leukaemia - A case report.

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Abstract:

Granulocytic sarcoma (chloromas) is a rare tumour usually associated with myelogenous leukemia. Tumour usually occur in childhood. Tumour is capable of invading the brain meninges or parenchyma. This case describes magnetic resonance imaging and Computed tomography appearance of intracranial chloroma. Chloroma shows some affinity for posterior fossa. On magnetic resonance imaging, lesion appears isointense on T1 weighted images, hypointense on T2 weighted images and Fluid-attenuated inversion recovery images with intense post contrast enhancement. There also can be bone remodelling seen.

Key Word: - Brain, Chloroma, Leukaemia, Myeloid sarcoma.

Introduction:

Granulocytic sarcoma or chloromas is a localized solid tumour composed of immature cells of the granulocytic series. Tumour has myelogenous origin associated with acute and chronic myelocytic leukemia. The majority of cases have association with acute myeloid leukemia (AML). Tumour occur mostly in childhood and frequently noted only at autopsy [1,12]. Granulocytic sarcoma may be seen in three clinical situations. First, they have association with known acute myelogenous leukemia (AML) [10]. Second, they may occur as an only finding in patients without a known diagnosis of leukemia. Finally, granulocytic sarcomas can occur in patients with myelodysplastic syndromes.

Granulocytic sarcomas are most commonly found in bone, periosteum, soft tissues, lymph nodes and skin. Intracranial involvement of the central nervous system by granulocytic sarcoma is rare[2,3].

This case report describes the magnetic resonance imaging (MRI) and computed tomography (CT) appearance of intracranial chloromas in a patient with acute myeloblastic leukemia.

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Case Report:

A 28-year-old man, previously in good health had fever, headache and bodyache 15 months ago. Evaluation by a general practitioner was done.

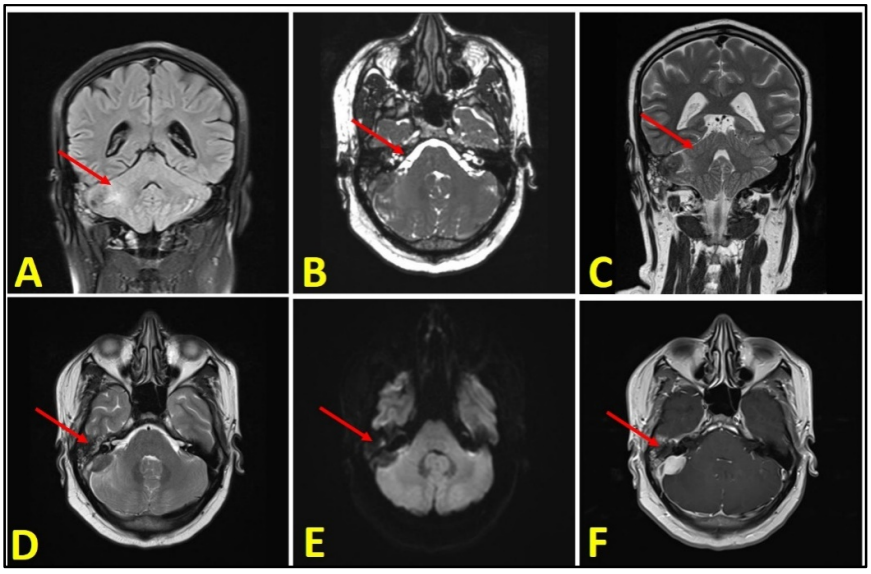
Routine blood investigation was performed.

- Hemoglobin 11.1 g/dl
- WBC count 31,700 cells/µl
- RBC count 5.28x10^6/cmm.
- Platelets 93x10^3 cells/cmm
- MCV 66.3 fl
- MCH 21.1 pg
- MCHC 31.8 g/dl
- RDW 15.9%
- polymorphs 85.3%
- lymphocytes 10.1%

On physical examination, patient was afebrile with pulse rate 86/min, respiratory rate 24/min, blood pressure 120/80 mmhg. Chest, CVS and CNS finding appears normal.

Patient was admitted to the hospital for evaluation. Bone marrow biopsy was performed. Immunophenotyping showed acute myeloblastic leukemia which is positive for AML1-ETO fusion and expressed myeloid markers like PO, CD15, CD13, CD33 and CD117 along with CD34 and HLADR. Patient was on chemotheraphy (Inj cytarabine 100 mg, Inj daunorubicin 20 mg and inj. filgrastrim 300 mcg) for 5 months and radiotherapy (24 Gy in 12 sittings). Suddenly patient developed right seventh-nerve palsy, and the high resolution CT of the head showed mastoiditis and hypodense lesion along right sigmoid sinus. Examination revealed, a right seventh-nerve palsy, and hepatosplenomegaly. The results of laboratory studies and another bone marrow biopsy supported previous findings. A MR scan of the brain was obtained 1 week after admission.

Image 1: MRI of Intracranial chloromas.



A. coronal FLAIR, B. CISS, C. coronal T2W, D. axial T2W, E. DWI, F. Post contrast T1W

(FLAIR= Fluid-attenuated inversion recovery, CISS= constructive interference in steady state, , T2W= T2 weighted, DWI= diffusion weighted image, T1W= T1W weighted)

In coronal FLAIR brain MR image, shows hypointense lesion along right sigmoid sinus infiltrating right cerebellum and shows perilesional oedema (arrow). (Image 1.A) CISS MR Image, shows lesion is far from intracranial and intracanalicular right facial

nerve also there is changes of right sided mastoiditis.(Image 1.B) Coronal T2W and axial T2W MR Images, shows well-defined extra-axial hypointense lesion along right sigmoid sinus with changes of mastoiditis.(Image 1.C & D) DWI MR Image shows, no restriction of above mentioned lesion.(Image 1.E) Post contrast T1W MR Image, shows intense homogenous post contrast enhancement.(Image 1.F) Single voxel spectroscopy MR image, shows increase choline peak in centre of lesion. (Image 2) Perfusion cerebral blood flow MR Image, shows decrease rCBF value suggest, hypo-perfusion of chloroma. (Image 3)

On the basis of the clinical setting and the CT and MR findings, the intracranial lesion were thought to be chloroma, and the patient had cranial irradiation. However, after irradiation patient lost to follow up so follow up investigation not done.

Image 2: Spectroscopy of Intracranial chloromas.

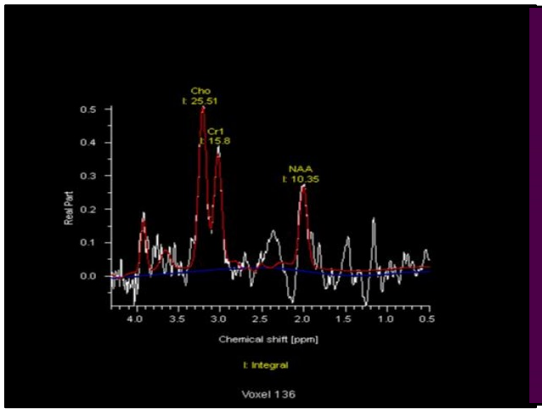


Image 3: Perfusion MRI of Intracranial chloromas.

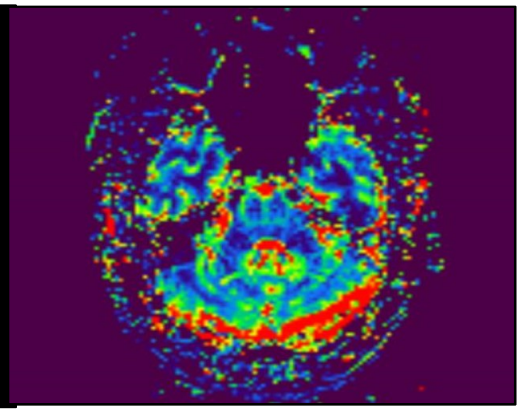
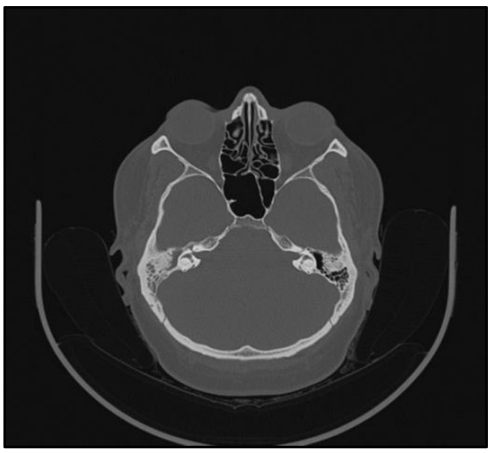


Image 4: CT image with bone window at same level shows no erosion of mastoid bone with smooth indentation due to lesion.



Discussion:

In 1853 [4] King described the term "chloroma" because of its characteristic green colour of tumour, which later was shown to because of the presence of myeloperoxidase in the tumour cells. Not all chloromas are green. Other synonyms are granulocytic sarcoma and myeloblastoma. The prevalence of chloromas is more in acute myelogenous leukemia,

however a few cases with chronic myeloproliferative disorders have also been described [3.]

Chloromas may be present at the time of diagnosis, or may precede the diagnosis by months to many years. Chloromas usually are found in association with bone; chloroma is thought to arise in the bone marrow and traverse the haversian canals to reach the periosteum. These tumors have shown a propensity for the cranium and facial bones and usually are attached to the dura or the periosteum of the bones of the paranasal sinuses, mastoid cells, or orbits [5]. Chloromas involving the CNS are exceedingly rare. In post-mortem studies, diffuse leukemic meningeal or dural infiltration is found in 25-50% of patients with leukemia [7]. Whereas Nodular infiltrates or parenchymal masses, have been reported rarely [5]. The advent of more effective chemotherapy has resulted in increased survival of patients with acute myelogenous leukemia, and consequently, the prevalence of CNS leukemia has increased. Azzarelli and Roessman[6] have reported that CNS leukemia is the result of the passage of neoplastic cells from involved bone marrow to the dura of the skull. Then, the cells by way of perivenous adventitial tissue connecting dura matter with subarachnoid space, can invade the brain parenchyma. On CT, intracranial chloromas may exhibit intermediate or high attenuation on unenhanced scans, may be associated with surrounding oedema, and typically show uniform contrast enhancement [3]. Differentials of this lesion are hematoma, abscess, meningioma, metastatic neuroblastoma, and CNS lymphoma [2, 11]. The hyperdensity, uniform enhancement, and surrounding oedema of this patient's intracranial lesions are certainly compatible with description of the CT appearance of intracranial chloromas. On MR images, the lesions were isointense with white matter on both the T1- and T2-weighted scans. Although this is an uncommon finding with intracranial masses, it has been described with meningioma. Kao et al. [9] reported an identical pattern in two other patients with chloromas. No comment was made about bone marrow, but the published images suggest abnormal marrow signal intensity. On the basis of one proposed pathogenesis of chloromas, we would expect this to reflect the intracranial extension of marrow infiltration. It is important to consider chloromas in the differential diagnosis of extraaxial intracranial masses. Although these lesions might be mistaken for meningiomas on the basis of their signal intensities on MR images, the appearance of abnormal bone marrow should suggest the correct diagnosis. MRI is preferred over CT for marrow imaging and for diagnosis of marrow involvement. We anticipate that MR contrast agents will increase the ability to detect these low-contrast lesions.

Conclusion:

Intracranial chloroma in case of leukemia is rare entity. Main differentials for intracranial extraaxial lesions are hematoma, abscess, meningioma, In view of known case of leukemia, it is important to consider chloromas in the differential diagnosis.

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