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CONCURRENT INTRACRANIAL CHLOROMAS AND LEUKEMIA CUTIS IN CHRONIC MYELOID LEUKEMIA

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ABSTRACT

Granulocytic sarcoma (chloroma) is an extramedullary solid tumor composed of immature white blood cells. Most common involvement sites are bone, periostium, soft tissue, lymph nodes and skin. Intracranial granulocytic sarcoma rarely occurs in meningeal or parenchymal form1. We report the of concurrent occurrence of intracranial chloromas and leukemia cutis in 26 years old male patient, known case of Chronic Myeloid Leukemia for 3 years [+ve Philadelphia (Ph) chromosome in bone marrow cells] who now presented to the Civil Hospital Karachi with continuous headache and multiple cutaneous nodules all over the body for last 2 months. The CT exam of the Head revealed multiple hyper dense extra-axial nodular masses (with mean value of 48 HU) which showed vivid enhancement on post contrast images, along with multiple subcutaneous enhancing nodules seen in the scalp and maxillofacial region. The CSF cytological examination confirmed presence of immature leukemic cells in the subarachnoid space. The subcutaneous chest nodule histopathology showed granulocytic sarcoma. To the best of authors' knowledge, the concurrent occurrence of granulocytic sarcomas at different locations in a single patient with Chronic Myeloid Leukemia has not been reported earlier in the literature.

Key words

Chronic Myeloid Leukemia; CML; CT; HU; leukemia cutis; intracranial chloroma; Granulocytic sarcoma.

INTRODUCTION

Chronic myelogenous (or myeloid or myelocytic) leukemia (CML) is a cancer of the white blood cells. It is a form of leukemia characterized by the increased and unregulated growth of predominantly myeloid cells in the bone marrow and the accumulation of these cells in the blood. It is a type of myeloproliferative disease associated with a characteristic chromosomal translocation called the Philadelphia chromosome. CML is now largely treated with tyrosine kinase inhibitors (TKIs) –imatinib which have led to dramatically improved long-term survival rates. CML accounts for 15-20% of all adult leukemia. Information on CML incidence and prevalence is scare, as CML is a rare disease [2]. CML is often divided into three phases based on clinical characteristics and laboratory findings. It typically begins in the chronic phase, and over the course of several years progresses to an accelerated phase and ultimately to a blast crisis. Blast crisis clinically behaves like an acute leukemia. Blast crisis is diagnosed if any of the following are present in a patient with CML: >20% myeloblasts or lymphoblasts in the blood or bone marrow or the Development of a chloroma/ Granulocytic sarcoma[3-4]. Granulocytic sarcoma is a extramedullary proliferation of leukemic cells. It a manifestation of acute myeloid leukemia (AML), chronic myeloid leukemia and other myeloproliferative disorders. They occur in 0.5-1.8% of patients with chronic myelogenous leukemia[5]. Most common involvement sites are bone, periostium, soft tissue, lymph nodes and skin. Intracranial granulocytic sarcoma rarely occurs in meningeal or parenchymal form[1]. CT and MRI have been the most commonly utilized imaging modalities for assessment of CNS myeloid sarcoma. These sarcomas are very sensitive to focal irradiation or chemotherapy[5]. To the best of authors' knowledge, the concurrent occurrence of multiple cutaneous granulocytic sarcomas as well as multiple intracranial granulocytic sarcomas have not been reported earlier in the literature. We hereby present concurrent intracranial chloromas and leukemia cutis in chronic myeloid leukemia as the first case report.

CASE REPORT

A 26-year-old Asian male, resident of thatta presented to the Civil Hospital 3 years back in August 2013 with complaints of chronic fever, abdominal pain and swelling. Fever was continuous, low grade and not relived by antipyretics. There was no significant family history. He underwent karyotype analysis as well as bone marrow trephine biopsy from right iliac blade. Karyotyping showed reciprocal translocation of chromosome segment between chromosomes 9 and 22. The biopsy revealed Philadelphia (Ph) chromosome in bone marrow cells and was labeled as chronic myeloid leukemia in chronic phase He was kept on tab Gleevec (Imatinib mesylate) PO 400 mg per day. On which his symptoms were well controlled. Meanwhile, the patient was lost on follow up visits due to personal issues and now he presented again to the Civil Hospital in September 2016 with complaints of innumerable cutaneous nodules all over the body for last 2 months and continuous severe headache for 25 days, not relieved by anlagesics. There was no history of trauma. On admission, laboratory investigations showed hemoglobin 8 g/dl, mean corpuscular volume 86.9fl, platelet count 132 $\times$ 10^9/L and white blood cell count 134.6 $\times$ 10^9/L, with monocytes(raised) 12%, neutrophils 66% and lymphocytes 11%. The peripheral smear revealed promyelocytes, myeloblasts, myelocytes, metamyelocytes, nucleated red blood cells are seen with basophilia and Leukoerythoblastic picture. Ultrasound

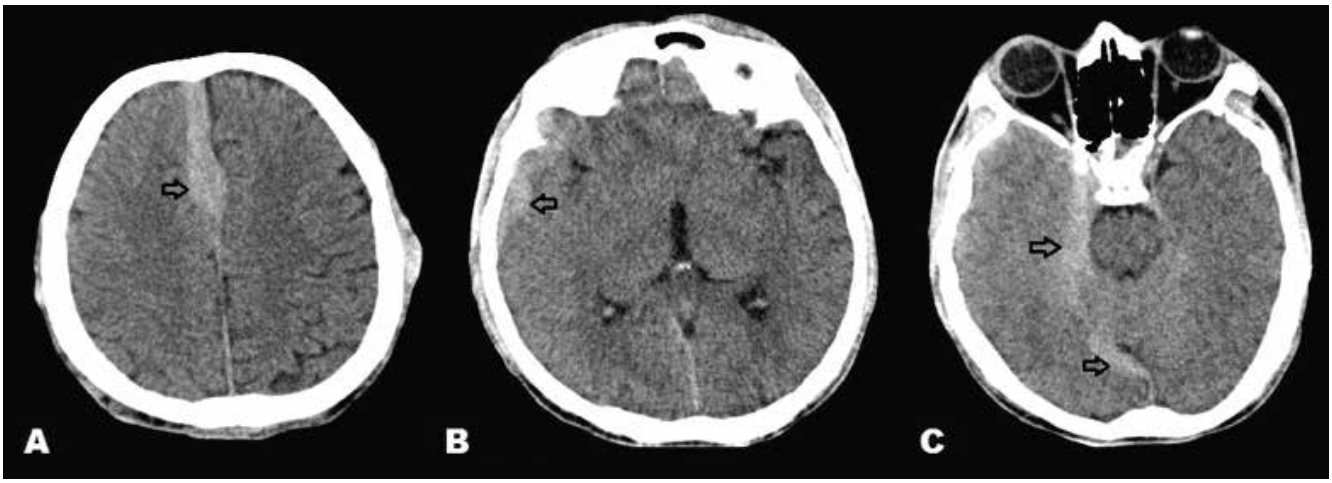

Figure: 1- Multi- axial Plain CT sections of brain above the level of lateral ventricles (A), at level of quadrigeminal plate cistern (B) and at the level of pons (C), showing hyper-dense intradural nodular deposits at parietal region, sylvian fissure and at para-falcine location along the tentorium cerebelli – [black arrows].

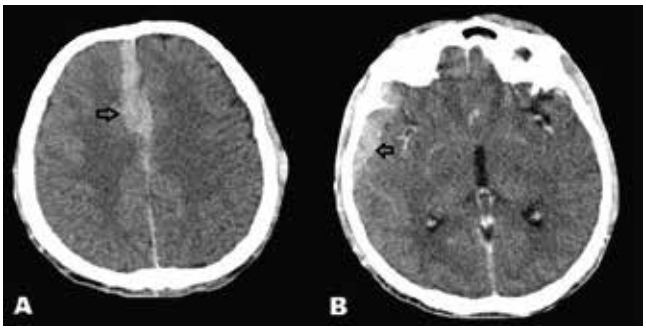

Figure: 2- Axial CT post contrast brain images showing enhancement of the extra axial lesions – [arrows in Fig. A & B].

exam revealed massive splenomegaly measuring 22.0cm. Computed tomography of the Head was performed on 16 slice Toshiba Activion Spiral CT scanner with dual head power injector and scanning parameters of 120 KV, 150mA and 5mm slice thickness; images were obtained in Pre and Post Contrast phases in multiple planes and viewed at appropriate window settings. There were multiple well defined intracranial confluent as well as discrete hyperdense extra-axial masses at right frontoparietal region, right sylvian fissure and at right para falcine region, also along the Tentorium cerebelli; Largest measures 8mm in width, they are hyper dense to gray

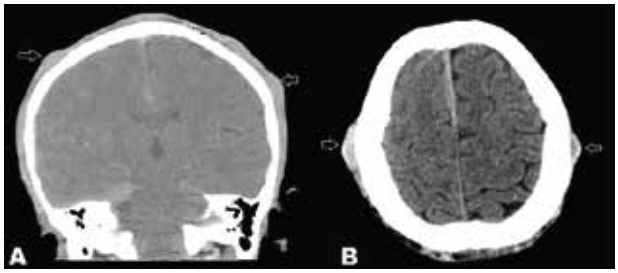

Figure: 3- Coronal (A) and Axial CT (B) images showing multiple soft tissue scalp subcutaneous enhancing nodules – [white arrows].

matter with mean value of 48 HU which demonstrated homogenous enhancement with post contrast mean attenuation value of 69HU. No adjacent parenchymal edema was seen. Bilateral orbital region was typically spared. Visualized bones appeared unremarkable. There were multiple subcutaneous enhancing nodules seen in scalp and maxillofacial region. Largest along the left maxillary labial fold measures 29x16mm. The CSF cytology via lumbar puncture confirms infiltration of myelocytes and myeloblasts in the subarachnoid space. The tru-cut biopsy of the largest subcutaneous nodule at left anterior chest wall measuring 40x40mm showed blast transformation to acute myeloid leukemia and diffuse infiltration of myeloid blast cells in the dermis and subcutaneous tissue.

DISCUSSION

Our case describes 26 years old male- a known case of Chronic Myeloid Leukemic for last 3 years, and was kept on glevaac and now presented with continuous severe headache and multiple nodules all over the body for last 2 months. Our clinical suspicion was brain leukemia. His CT of the head demonstrated the presence of multiple hyper dense enhancing intra cranial extra-axial nodular masses, confirmed as immature leukemic cells on CSF cytologic examination. The investigations revealed blast crises of CML and concurrent occurrence of granulocytic sarcoma in skin and intracranial space. O'Brien CE et al in 2011[6] at Washington, DC presented a case report of intracranial extra axial Granulocytic Sarcoma on CT in a34-year-old chronic myelogenous leukemic patient with blast crisis mimicking as subdural hematoma -comparable to our case. In 2015, Cervantes GM et al[7] studied summary of clinical findings, CT and MR features of intracranial myeloid sarcomas (MS) in 21 reported cases presenting with AML from 1971 to 2014. 12 out of a total of 24 lesions were assessed with CT. Among these, 11 lesions (91%) appeared hyperdense on non-contrast CT. Out of all 11 hyperdense lesions, six lesions were intra-axial and five extra-axial. 91% lesions exhibited avid homogeneous enhancement. The CT findings of these case reports were compatible to our case.

In 2011, Out of 60 patients of chronic myeloid leukaemia studied over 21 months at Rawalpindi by Ahmed S. et al [8], 6(10%) cases of granulocytic sarcoma were encountered, sites of extramedullary disease were skin, breast, spinal canal and lymph nodes and none had intracranial involvement. Kanade U et al[9] reported a case of chronic myelogenous leukemia cutis in a 70-year-old male patient presented with nonspecific abdominal discomfort and multiple papular and nodular skin lesions all over the body for 6 months. Aspiration cytology smears from cutaneous nodule revealed variable mixture of mature and immature cells of granulocytic series – again comparable to our case. To the best of authors' knowledge, the concurrent occurrence of multiple cutaneous granulocytic sarcomas as well as multiple intracranial granulocytic sarcomas in chronic myelogenous leukemia have not been reported earlier in the literature. The patient was referred to the oncology department for AML induction chemotherapy.

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Ateeque Ahmed Khan; Study concept and design, protocol writing, data collection, data analysis, manuscript writing, manuscript review
Mahnoor Hafeez; Study concept and design, data collection, data analysis, manuscript writing, manuscript review