Zuckerguss Sign: Sugar-Coated Cerebellum in a Case of Gastric Adenocarcinoma

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A 45-year-old female patient was brought to the emergency department with a history of headache since 2 weeks, slurring of speech, and unsteadiness of gait since 1 week. She developed one episode of generalized tonic-clonic seizure 1 day before. She was diagnosed with adenocarcinoma of the stomach 6 weeks before, however deferred treatment.

She was emaciated with stable vitals. On neurological examination, she was conscious, alert, and oriented. The findings of the fundus examination were normal with both pupils normal size and reacting to light. She had ataxic dysarthria, dysmetria, and gait was broad-based with impaired tandem walking. Terminal neck stiffness was observed.

Routine laboratory investigations showed the hemoglobin of 9 gr/dL, random blood sugar of 96 mg/dL, and serum creatinine of 0.6 mg/dL. Serum electrolytes, liver function tests, serum calcium, and thyroid profile were normal and HIV 1 and 2 was non-reactive. She was further evaluated with magnetic resonance imaging (MRI) of brain plain and contrast, which revealed diffuse, thick sheet-like leptomeningeal enhancement seen along bilateral cerebellar folia giving sugar-coated appearance [Figure 1].

Cerebrospinal fluid (CSF) analysis was abnormal with a protein of 60 mg/dL, sugar 65 mg/dL, and cell count of 25/cumm. Meningoencephalitis panel was negative for infective etiology.

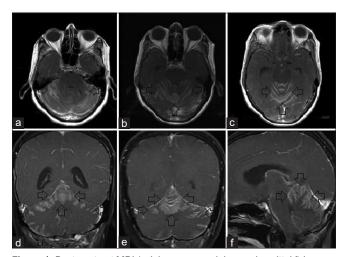


Figure 1: Post contrast MRI (axial a–c, coronal d, e, and sagittal f) images of brain show thick sheet-like leptomeningeal enhancement (black open arrows) seen along cerebellar folia giving sugar-coated appearance

Cytology revealed abnormal clusters of large atypical cells suggestive of malignancy [Figure 2].

Review of previously done gastric biopsy slides depicted sheets of atypical cells, few with signet ring morphology suggestive of adenocarcinoma [Figure 3]. The final diagnosis of carcinomatous meningoencephalitis with classical Zuckerguss or sugar-coated appearance on MRI brain in a known case of adenocarcinoma stomach was made. She was started on intravenous levetiracetam 1500 mg loading followed by continuation of 500 mg twice a day and intrathecal twice-weekly triple-drug therapy with a combination of methotrexate, hydrocortisone, and cytarabine. Over the next 4 weeks, she was symptomatically better but lost to further follow-ups.

Çarcinomatous meningitis usually occurs from solid tumors such as carcinoma lung, breast, and melanoma.^[1] Common clinical manifestations include subacute onset of headache, encephalopathy, seizures, and focal neurological deficits. Primary gastric adenocarcinoma resulting in carcinomatous meningitis is extremely rare and can occur as a manifestation of relapse also.^[2] Repeated CSF cytospins may be required for establishing the diagnosis.

Imaging features of carcinomatous meningitis include diffuse or scattered meningeal enhancement with or without hydrocephalus. Extensive posterior fossa involvement with enhancement of leptomeningeal spaces of cerebellar foliae and brainstem can give the sugar-coated appearance or Zuckerguss pattern.^[3] In the appropriate clinical context, this imaging

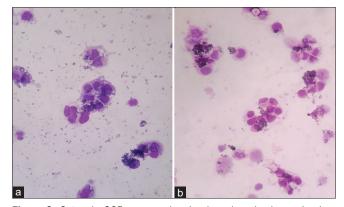


Figure 2: Cytospin CSF smears showing loosely cohesive and acinar clusters of large atypical cells having hyperchromatic, pleomorphic nuclei, suggesting metastatic adenocarcinoma (a: MGG stain, b: H and E stain)

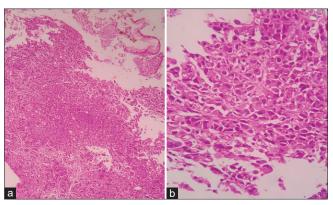


Figure 3: Stomach biopsy showing sheets of large atypical cells having hyperchromatic, pleomorphic nuclei. Some cells show signet ring cell morphology (a: low power and b: high power)

finding is highly suggestive of carcinomatous meningitis, which should alert the clinicians for early diagnosis and plan proper management protocols.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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Submitted: 10-Jun-2021 Revised: 01-Sep-2021 Accepted: 20-Sep-2021

Published: 15-Feb-2022

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DOI: 10.4103/aian.aian_535_21