ORIGINAL ARTICLE

Milky strawberry-like cerebrospinal fluid secondary to hypertriglyceridemia. A rare case report of hemorrhagic stroke

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Abstract:

Introduction: Disorders of lipid metabolism can lead to intracranial hemorrhages as a rare complication of the disease. To our knowledge only 4 case reports were found to have milky pinkish cerebrospinal fluid secondary to severe hypertriglyceridemia.

Case description: 40-years old Indian male, presented to the Emergency Department with severe headache, vomiting and decreased level of consciousness. His CT brain showed intraventricular hemorrhage causing secondary hydrocephalus. His lab investigations revealed very high lipid profile specially triglycerides level (70.19 mmol/L). An external ventricular drain (EVD) was inserted and cerebrospinal fluid was milky strawberry-like. Cerebrospinal fluid analysis revealed high lipid profile as well. Both CTA and DSA were unremarkable for any vascular malformations. The patient was successfully managed through a combined effort of Neuro-Surgery, medical and ICU teams. His serum lipid profile came back to normal, his external ventricular drain was removed and a ventriculo-perotineal shunt was inserted.

Conclusion: The case illustrates the significant high risk for intracranial hemorrhages secondary to hyperlipidemia. Therefore, it may be suggested to obtain a cerebrospinal fluid sample via lumbar puncture for example to measure the corresponding cerebrospinal fluid lipid profile in patients with extremely high serum levels. This may possibly predict the associated high risk for these devastating intracranial hemorrhages. The guidelines for the serum level at which this has to be performed should be tailored based on multiple randomized multicenter clinical trials. The combined effort of the medical, Neuro-Surgical and ICU teams is the cornerstone in the management of these special cases.

Keywords: Milky cerebrospinal fluid, hypertriglyceridemia, hemorrhagic stroke, ventriculo-perotineal shunt

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Introduction:

Disorders of lipid metabolism can have adverse effect on the whole-body systems, homeostasis and hemostasis. These disorders may lead to complications like diabetes, hypertension, cerebrovascular disorders, myocardial infarction, venous thromboembolisms and postpartum hemorrhage.[1] Their effect may progress furthermore to affect the fetal outcomes in pregnant ladies resulting in premature births, increased risks for fetal congenital anomalies as well as growth restriction.[2]

Intracranial hemorrhages secondary to hyperlipidemia is a rare complication of the disease and is associated with poor prognosis and outcome.[3,4] To our knowledge only 4 case reports were found to have milky strawberry cerebrospinal fluid secondary to severe hypertriglyceridemia one of them died soon after surgical intervention despite the aggressive supportive management and the others required a combined effort of medical team, ICU team and neuro-surgery team.[5,6]

In hyperlipidemia, the tendency to bleed is not only confined to the brain. In other words, beside intracranial hemorrhages the patient may

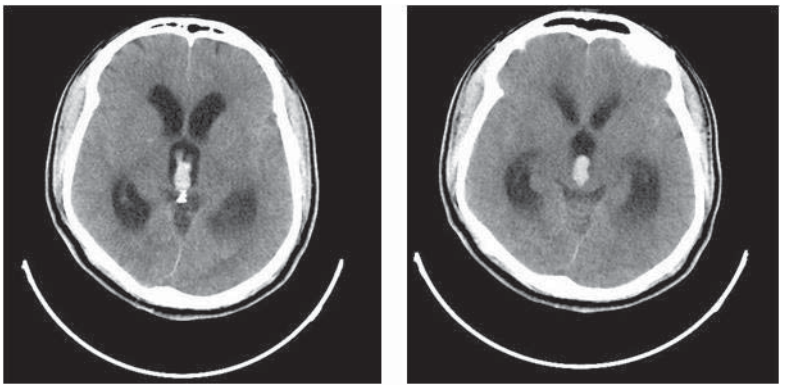

Figure 1: The patient's CT brain showing IVH in the third ventricle and occipital horns of the lateral ventricles with significantly dilated ventricles

Figure 2: The drained CSF showed milky strawberry-like color with thicker than usual CSF consistency.

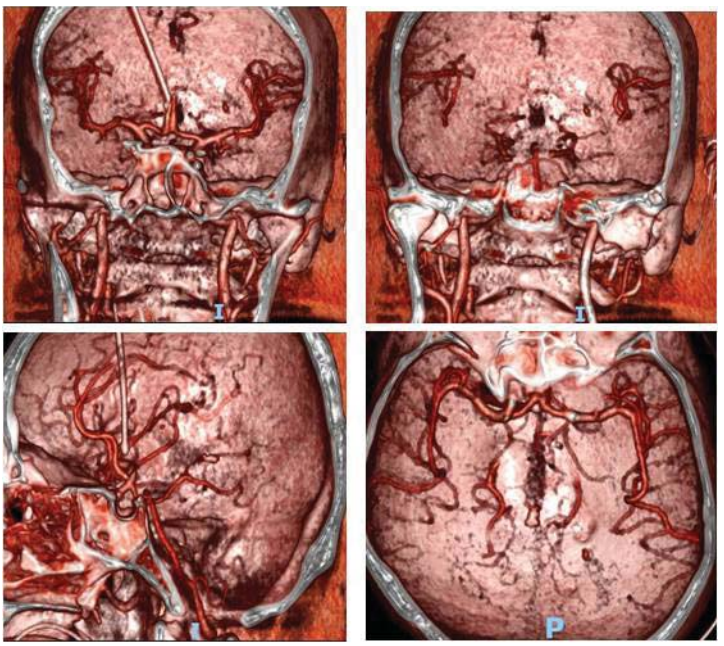

Figure 3: Showing normal CTA 3D, no aneurysm seen.

have postpartum hemorrhage, Sheehan's syndrome, pituitary crisis as well as other many hemorrhagic disorders.[1]

In this study, we would like to report a rare case of milky strawberry-like cerebrospinal fluid secondary to hypertriglyceridemia who presented with a hemorrhagic stroke in the form of spontaneous intraventricular hemorrhage.

Case description:
40-years old Indian male known diabetic and hypertensive for more than 20 years, presented to the emergency department with severe headache and episode of vomiting with decreased level of consciousness. Upon arrival to emergency department, he was intubated and sedated by the ER physician.

On examination GCS was 7T/15 E2VtM5 pupils were 2mm sluggishly reactive to light, moving both sides adequately to painful stimuli. No tendon xanthoma, xanthelasmata, corneal arcus or any other signs of hyperlipidemia were detected.

Computed tomography (CT) brain showed intraventricular hemorrhage (IVH) causing secondary hydrocephalus with small bilateral parieto-temporal subarachnoid haemorrhage. (Fig.1)

Laboratory results revealed blood glucose of 6.21 mmol/L (normal range "NR": 4.11–5.89), total cholesterol of 34.8 mmol/L (NR: 0.9–5.2) low-density lipoprotein of 255 mg/dL(NR: 130 -159), high-density lipoprotein of 0.27 mmol/L (NR: 1.04 – 1.55) and triglycerides of 70.19 mmol/L (NR: 0.1 – 1.7).

Therefore, the patient was prepared and taken to the OR where an emergent external ventricle drain (EVD) was inserted. The drained cerebrospinal fluid (CSF) was surprisingly milky strawberry-like to the degree it was expected to be mixed with pus for its unusual color and consistency. (Fig.2)

CSF samples were sent and showed W.B.C 0.022 X10^3/UL, Glucose 11.12 mmol/L (NR: 2.2 – 3.9), protein 95.5 mg/Dl (NR: 150 – 450),

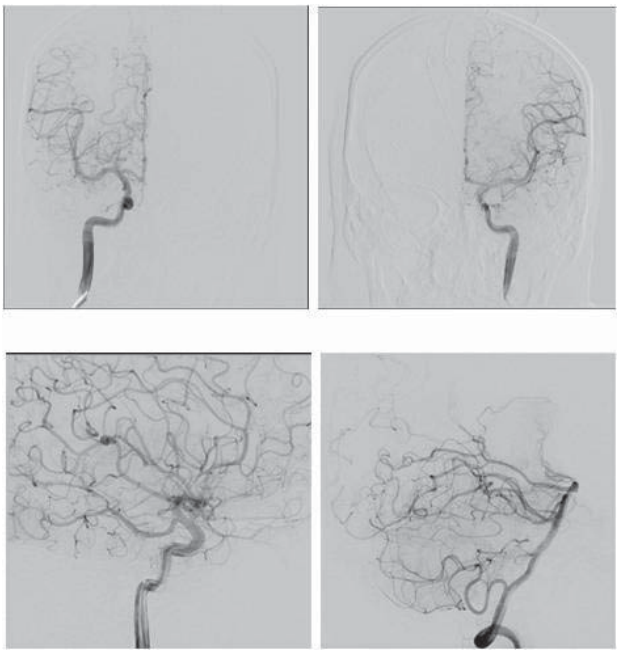

Figure 4: DSA with AP and Lateral views of circle of willis, showing no detectable vascular anomaly.

Figure 5: Showing significant change in the CSF color and consistency after plasma electrophoresis and lipid-lowering medications

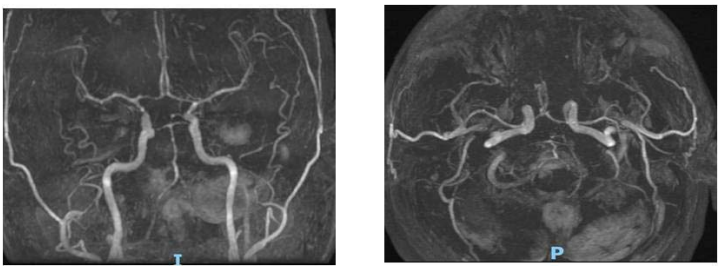

Figure 6: Showing small caliber of bilateral distal vertebral arteries, basilar artery and bilateral PCA, which indicates significant vasospasm

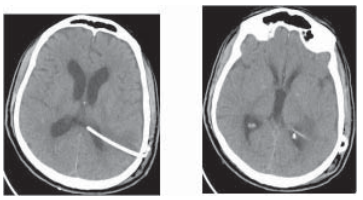

Figure 7: Showing latest patient's CT brain, showing placement of the VP shunt and resolving IVH

LDH CSF 59 U/L (NR: 0 – 10), triglycerides CSF < 0.11 (NR: < 1.7).

Initially brain CT angiography done to exclude any aneurysms or vascular malformations, but it was inconclusive (figure.3), therefore a definitive DSA was done and confirmed the absence of any detectable vascular malformations as shown in figure 4.

The patient was diagnosed as a case of hemorrhagic stroke secondary to hypertriglyceridemia- related milky strawberry-like cerebrospinal fluid. He was started on Fenofibrate 145mg daily for 2-weeks and underwent 3-sessions of plasma electrophoresis. After that both cerebrospinal fluid color and consistency significantly improved as shown in figure-5.

After that, he improved gradually, GCS increased to 11/15 E4V1M6, he was successfully extubated, EVD removed, but eventually he required a permanent ventriculo-perotineal shunt. (figure-7)

His latest lipid profile was triglycerides 2.25 mmol/L (NR: 0.1 – 1.7), HDL 0.46 mmol/L (NR: 1.04 – 1.55), cholesterol 2.15 mmol/L (NR: 0.9 – 5.2) and LDL of 0.66 mg/dL (NR: 130 -159).

Conclusion:

The case illustrates the significant high risk for intracranial hemorrhages secondary to hyperlipidemia. This is especially seen when the serum levels are unexpectantly very high. Therefore, it may be suggested to obtain a cerebrospinal fluid sample via lumbar puncture for example to measure the corresponding cerebrospinal fluid lipid profile in patients with extremely high serum levels. This may possibly predict the associated high risk for these devastating intracranial hemorrhages. Further more, it will allow aggressively and rapidly lowering the serum levels; through plasma electrophoresis for example or any other measure; before hand. The guidelines for the serum level at which this has to be performed should be tailored based on multiple randomized multicenter clinical trials.

The collaboration between the medical, neurosurgical and ICU teams is the corner stone in the management of these special cases.

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Role and contribution of authors:
Mohammed Awad Elzain, the conception and design of the work, writing the article including the literature review and the conclusion.

Mostafa Osman, the conception of the work, revising the article critically for important intellectual content and final approval of the version to be published.

Mohammed N. Al-Othman, participated in writing the case description section and revising the article critically for important intellectual content and final approval of the version to be published.

Rabab Abdul Karim T Boukhari, participated in writing the case description section and revising the article critically for important intellectual content and final approval of the version to be published.

Zaghloul Mahmoud Moussa, revising the article critically for important intellectual content and final approval of the version to be published, besides operating on the patient as the primary surgeon.

Saad Al-Shahrani, revising the article critically for important intellectual content and final approval of the version to be published, besides being the responsible attending consultant managing the case.

References:

1. Liu DS, Liu L, Gan F, Wu XL, Ye G. Acute Pancreatitis Complicated by Sheehan's Syndrome: A Case Report and Literature Review. Chin Med Sci J 2020;35:95-100.
2. Cai E, Czuzoj-Shulman N, Malhame I, Abenhaim HA. Maternal and neonatal outcomes in women with disorders of lipid metabolism. J Perinat Med 2021;49:1129-1134.
3. Noda H, Iso H, Irie F et al. Low-density lipoprotein cholesterol concentrations and death due to intraparenchymal hemorrhage: the Ibaraki Prefectural Health Study. Circulation 2009;119:2136-2145.
4. Wang X, Kong Y, Chen H, Zhang JH, Wang Y. Characteristics of acute cerebral hemorrhage with regard to lipid metabolism and glycometabolism among different age groups. Acta Neurochir Suppl 2011;111:367-371.
5. H.-W.Chen, Lotung, Yi-Lan, Taiwan, S.W.Lin, C.-C.Lai. Milky cerebrospinal fluid and serum in hypertriglyceridemia. [108], 77-78. 6-30-2014. Q J Med. 1-1-2015. Ref Type: Case
6. Su Jiahao, Ba Yueyang, Liang Sitao, Liu Huijiao. High-Lipid Cerebrospinal Fluid: A Case Report. Neurology India 2020;68:525-527.