Delirium Precipitated by Polycythaemia

Introduction

Delirium is a common and serious problem among acutely unwell persons. Although linked to higher rates of mortality, institutionalization and dementia, it remains under diagnosed. Careful consideration of its phenomenology is warranted to improve detection and therefore mitigate some of its clinical impact. The publication of the fifth edition of the Diagnostic and Statistical Manual of the American Psychiatric Association (DSM-5) provides an opportunity to examine the constructs underlyng delirium as a clinical entity [1]. We are reporting a case of delirium in a 20 years old female that was precipitated by a secondary polycythaemia caused by a congenital cardiac left to right shunt resulting in a pulmonary hypertension.

Our med pub mesh literature search did not yield a previously reported similar case.

As far as we know we are reporting a first case of its kind.

DSM classifications of delirium

a. Disturbance in attention (reduced ability to direct, focus, sustain, and shift attention) and awareness (reduced orientation to the environment).

b. The disturbance develops over a short period of time (usually hours to a few days), represents a change from baseline attention and awareness, and tends to fluctuate in severity during the course of a day.

c. An additional disturbance in cognition (e.g. Memory deficit, disorientation), language, visuospatial ability, or perception that is not better explained by a preexisting, established, or other evolving neurocognitive disorder.

d. The disturbances in Criteria A and C are not better explained by another preexisting, established, or evolving neurocognitive disorder and do not occur in the context of a severely reduced level of arousal, such as coma.

e. There is evidence from the history, physical examination, or laboratory findings that the disturbance is caused by the physiological consequence of another medical condition, substance intoxication or withdrawal (i.e., due to a drug of abuse or to a medication), or a toxin exposure, or is due to multiple etiologies.

While there was no denial of the organic etiology of delirium, based on a huge body of references that surpassed million hits related to the subject. The etiologies of delirium are diverse and multifactorial and often reflect the pathophysiologival consequences of an acute medical illness, medical complication or drug intoxication. Delirium can have a widely variable presentation, and is often missed and under diagnosed as a result [2].

Polycythaemia and delirium

It was reported in numerous papers that Polycythaemia Vera has been a regular but not so common etiology of delirium as reported by Polycythaemia, delirium and mania [3]. But our review of literature yield much less reports of Delirium precipitated by secondary Polycythaemia, found in Erythremia (polycythemia) with a psychosis erythremia was one of the factors responsible for the appearance of the psychosis [4]. Here we are presenting the case of 20 years old, new bride who arrived from his country of origin 3 weeks before admission to the Hospital. The marriage was arranged and the family of the husband has no past history of the woman mental or physical status. They presented to the Emergency Room with her family due to change in her behavior and a consistent headache for the last one week. The Patient was fearful, anxious and at times confused, disoriented to time, place and person.

Psychiatry was consulted and the initial evaluation reported that she didn’t show any sign or symptom of mood or psychotic disorder during the interview. She was with poor concentration, poor attention span, forgetful. She was having episodes of confusion, disorientation and incoherence with fluctuations.

Prior to her presentation to Emergency her family took her to a traditional healer which left superficial marks on her scalp with no improvement. The Psychiatrist on call cleared her from their side and recommended to have Medical consultation. The initial basic blood investigation showed an unexplained and serious elevation in her hematocrit (70) and Hemoglobine (18’7), MCV (75’8). So she was admitted to the Medical ward for further investigation where a Hematologist, a Cardiologist and a Neurologist were consulted.
During admission 2 venesections were done, secondary causes of Polycythemia were investigated, pulmonary hypertension was found and other causes such as Viral infections and renal diseases were excluded. CT venogram, CT head and Chest X-ray were normal. Echocardiography showed a severe pulmonary hypertension, EF 55-60 %, right ventricle was moderately enlarged, the right ventricular systolic function was moderately impaired. She was diagnosed as Delirium secondary to general medical condition, Polycythemia secondary to pulmonary hypertension (due to a congenital right to left heart shunt). She was further hospitalized for four days, treated with one mg of haloperidol twice a day and exsanguinations as ordered by medicine; her orientation improved and was discharged with outpatient dini appointment for further investigation of possible cause of Pulmonary Hypertension due to Congenital Heart disease. She was started on low dose Haloperidol that helped but the dramatic response was with exsanguinotransfusion. Improvement in alertness correlated very well with the increase in cerebral blood flow which followed venesection [5].

Conclusion

We need to be aware that a secondary polycythemia can cause delirium such as found in heavy smokers, chronic COPD patients and residents of high elevations mountains, as presented in Mountain sickness [6]. Smoking as a cause of erythrocytosis [7]. The management of Polycythemia secondary to pulmonary hypertension needs to be multidisciplinary due to several possible etiologies; thus an inclusive differential diagnosis would be prudent standard of care in cases of delirium in non risk prone population.

References

1. European Delirium Association and American Delirium Society (2014) The DSM-5 criteria, level of arousal and delirium diagnosis: inclusiveness is safer. BMC Medicine 12: 141.
2. Tamara G Fong, Samir R Tulebaev, Sharon K Inouye (2009) Delirium in elderly adults: diagnosis, prevention and treatment. Nat Rev Neurol 5(4): 210-220.
3. Chawla M, Lindesay J (1993) Polycythemia, delirium and mania. Br J Psychiatry162: 833-835.
4. Levin M (1938) Erythremia (polycythemia) with a psychosis. Am J Psychiatry 10: 407-410.
5. Willison JR, Thomas DJ, du Boulay GH, Marshall J, Paul EA, et al. (1980) Effect of high haematocrit on alertness. Lancet 1(8173): 846-848.
6. Buldas J (2015) Mountain sickness. CasLekCesk 154(6): 280-286.
7. J Moore-Gillon (1975) Smoking as a cause of erythrocytosis. Ann Intern Med 82(4): 512-515.