Bilateral Central Vein Occlusion in a Case of Diaphragmatic Eventration

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ABSTRACT
Central Retinal Vein Occlusion (CRVO) is a severe retinal pathology, which causes visual impairment usually after the age of 40. Mostly unilateral, less than 10% of cases are bilateral. Affected young adults (under the age of 40), usually exhibit a hidden, underlying systemic disease. Thorough testing has to be done in order to spot the pathogenic agent. We present the case of a 25 years old woman with bilateral CRVO caused by Diaphragmatic Eventration and Chronic Respiratory Failure.

Keywords: central retinal vein occlusion, diaphragmatic eventration, chronic respiratory failure, obesity

INTRODUCTION
Central Retinal Vein Occlusion is a common retinal vascular disorder, with potentially painful and blinding consequences. Although it can occur at almost any age, it usually affects patients passed their fourth or fifth decade (1) with a history of diabetes, hypertension, hypercholesterolemia, obesity or smoking. Only 10-15% of CRVO patients (2) are younger than 40 and lack the previously mentioned risk factors. These require detailed examination, including medical history, clinical evaluation and specific investigations to elucidate the pathogenic cascade and prevent further damage.

CASE REPORT
We have the case of a 25-year-old woman admitted to our clinic for sudden, painless and consecutive visual loss in both eyes, accompanied by migraine. Her current complaints began almost 4 months ago with sudden and painless loss of visual acuity in her left eye (LE), which was soon followed by her right eye (RE). At that time, she was diagnosed as having Bilateral Papillitis and was admitted to a regional hospital, where she underwent systemic anti-
biotherapy and cortisone therapy. We have no data as to her visual acuity at that moment. The tests and examinations of the patient, that she was submitted to at that time, revealed multiple and heterogeneous pathologies: Right situated diaphragm? Elevated right hemidiaphragm (chest X-ray), type 2 Diabetes, Class II Obesity, Pickwick Syndrome, Sleep Apnoea Syndrome, Polycystic Ovarian Syndrome and Secondary Polyglobulia. Upon discharge, visual acuity in her right eye was 18/20 and hand motion perception was present in her left eye. Fundus examination revealed an ill defined right optic disc with surrounding, partially resorbed flame shaped haemorrhages as opposed to the congestive optic disc in her left eye with preapillary and peripapillary haemorrhages.

The patient’s status continued to worsen. At admittance to our health service she was confused and had difficulties expressing herself. Her recorded blood pressure was 150/100 mmHg, in lack of any treatment. Visual acuity was 20/400 in her right eye and no light perception in her left eye. Intraocular pressure was within normal limits. Pupillary light reflex was present and diminished in her RE, while LE presented a relative afferent papillary defect (RAPD). While anterior segment examination was unremarkable, fundoscopy revealed severe papillary oedema with drowning preapillary and peripapillary haemorrhages, macular star hard exudates, dilated tortuous retinal veins; small and medium sized retinal haemorrhages with peripheral extension (Figure 1A, B).

Laboratory tests revealed a non-specific inflammatory syndrome (Fibrinogen 488 mg/dl, CRP 6mg/dl ESR 5mm/h), high serum lipid levels (LDL cholesterol 189mg/dl, triglycerides 169 mg/dl) and normal values for serum proteins. RF, CIC, C3 and ASLO were within normal limits. Tests for ANA, c-ANCA, p-ANCA, aCL, LA were all negative. Evaluation of Protein C, Protein S, Antithrombine III levels and usual coagulation tests showed regular values. Liver and kidney function tests were also within normal range. Previously diagnosed polyglobulia, was confirmed by CPC and hematology consult (RBC 7.53 x 10^12 /L, Ht 60.9%, Hb 17.3 g/dl). A possible neurological condition was excluded after performing a head CT and a neurology consult. By means of endocrine evaluation and blood levels for cortisol, TSH and FT, we ruled out Myxedema and Cushing Syndrome. Further tests and evaluations were performed in order to assess the patient’s health. While some of these confirmed the previously mentioned diagnoses, others revealed newly found associated conditions as: Moderate Pulmonary Hypertension, Chronic Cor Pulmonale, Tricuspid Valve Insufficiency and Hypoxic Respiratory Failure. Subsequent chest CT indicated right diaphragmatic eventration and a pulmonary consolidation process at the right lobe medial bronchia (Figure 2).

Clinical diagnosis was obvious and it did not elicit any doubts. On the other hand, identifying the ethology of the pathogenic process was of utmost importance. Purtscher Retinopathy may share similarities with the current condition (raised thoracic pressure), but it is the peri-
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Maedica
A Journal of Clinical Medicine, Volume 9 No.3 2014

tive purposes. Open transthoracic diaphragm
plication was performed. Nevertheless, visual
prognosis was dramatic as her visual acuity was
very low.

DISCUSSION

Although CRVO is usually considered the
appanage of the elderly (6,7), it can arise in
young people lacking the common atheroscle-
rotic risk factors. Moreover, bilateral CRVO in
the young is a severe, life-altering event linked
to coagulopathies (8), dyspreoteinemias (9),
hyperhomocysteinemia (10,11), severe hyper-
tension (12,13), malignancies (14,15) or other
systemic disorders that requires immediate at-
tention (16).

To our knowledge, this is the first case of
bilateral CRVO following diaphragmatic even-
tration. Improperly called ‘diaphragmatic even-
tration’, the patient’s condition is a congenital
developmental defect in the muscular portion
of the diaphragm, as the normal attachments to
the sternum, ribs and dorsolumbar spine are
maintained3. More frequent in men, it is usu-
ally located on its left side and asymptomatic.

In our case, diaphragmatic eventration cau-
sed respiratory failure with secondary polyglo-
bulia. In addition, venous drainage was impa-
i red due to raised intra-thoracic pressure. At
the same time, the ongoing atherosclerotic process
was slowly disrupting vascular endothelium,
and thus enhancing circulatory difficulties. Any
of these could have caused the thrombosis by
itself. Atherosclerotic damage is widely recog-
nized as a prime factor of vascular conditions in
older patients. Complicated atherosclerosis
with CRVO or other ischemic events, imply ex-
tensive endothelial injury, developed over
many years. Hence, in our case, atherosclerosis
involvement was secondary. While polyglobu-
lia led to hyperviscosity with a consecutive
drop in local blood flow speed, the already
damaged endothelium further enhanced the
local circulation crisis. On the other hand, ve-
nous drainage was reduced due to the raised
intra-thoracic pressure and pulmonary hyper-
tension, ultimately leading to venous blood
stagnation and retinal vein thrombosis.

CONCLUSION

CRVO develops as the vile consequence of
systemic disorders. To prevent further dam-
ge, careful and complete examinations are
needed, acknowledging both common and un-
papilar haemorrhages and cotton wool spots
that clearly separate these disorders. Haemato-
logical disorders (leukaemia, anaemia) or co-
agulopathies causing a hyperviscosity syndrome
must be taken into account, considering the
patient’s young age and bilateral condition.
Except for polyglobulia, these were all ruled
out by CBC, Protein C and S levels and coagu-
lation tests. Waldenström macroglobulinemia
or Multiple myeloma was highly unlikely as se-
rum protein levels were normal. In addition,
antibody test results were negative for vasculi-
tis. (SLE, Churg-Strauss Syndrome, Wegener
granulomatosis, Antiphospholipid Syndrome).
Syphilis and HIV serology was also negative. At
the same time, head CT and neurology exam
ruled out the fiery possibility of a hypertensive
retinopathy, an intracranial haemorrhage, an-
eurysm or tumour. Albeit young, the patient
had already developed common, atheroscle-
rotic risk factors including diabetes, arterial hy-
pertension, hypercholesterolemia and obesity.

Final diagnosis for this patient was: Bilateral
Central Retinal Vein Occlusion. Right Dia-
aphragmatic Eventration. Hypoxic Respiratory
Failure. Chronic Cor Pumonale. Moderate Pul-
monary Hypertension. Secondary Polyglobulia.
Sleep Apnoea. Pickwick Syndrome. Metabolic
Syndrome. Grade II Hypertension. Tricuspid
Valve Insufficiency. Type 2 Diabetes.

According to literature guidelines (3-5), sur-
gical intervention was recommended for cura-
common risk factors. Adequate management of CRVO depends upon good cooperation with potentially associated specialties (neurology, endocrinology, internal medicine, laboratory medicine, pulmonology, radiology).

Conflict of interests: none declared.
Financial support: none declared.
Acknowledgement: The authors would like to thank Professor Dana Pelipceanu for writing assistance.

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