Case Number 5
Anton's Syndrome

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Case Summary:

Demographic details:

Mr. AS, male, Nadur
Referred from: Gozo General Hospital

57-year-old male admitted to ITU with a severe Community Acquired Pneumonia. Stayed in ITU for about three weeks and during this period suffered an episode of deterioration which was attributed to multiple venous infarcts of the brain. This caused the patient to become blind.

Presenting complaint:

Sudden onset blindness.

History of presenting complaint:

The blindness occurred suddenly and with no warning. Patient became increasingly disoriented and was clearly blind although adamantly kept saying that he was able to see normally. At the time the patient had been in ITU for about three weeks.

Past medical and surgical history:

Past medical history:

- Severe Community Acquired Pneumonia lasting three weeks, exactly preceding the blindness. Patient had to be admitted to ITU due to the severity of the pneumonia.
- Mild Hypertension.

Past surgical history:

- Patient did not have any major operations.

Drug history:

Drug	Dosage	Frequency	Type	Reason
Tazocin Piperacillin/Tazobactam	Piperacillin 4g Tazobactam 0.5g	TDS	Antibiotic	Treatment for pneumonia
Levofloxacin	500mg	Dly	Antibiotic	Treatment for pneumonia
Doxycycline	100mg	BD	Antibiotic	Treatment for pneumonia

Antibiotics were later switched due to the possibility of a viral pneumonia.

Drug	Dosage	Frequency	Type	Reason
Ganciclovir	300mg - IV	BD	Antiviral	Treatment for CMV pneumonia

Patient received IV ganciclovir for 12 days and was later switched to oral valganciclovir.

Family history:

The patient did not have any major illnesses in the family. His father had hypertension, as did his uncle but other than that nil of note.

Social history:

Patient used to smoke cigarettes and drink alcohol on occasions. Patient did not divulge the actual amount of cigarettes and alcohol he consumed.

Systemic inquiry:

- General Health: patient looks very thin and showed cachexia due to the long stay in ITU.
- Cardiovascular System: relatively normal except for mild hypertension.
- Respiratory System: patient was still short of breath even after the pneumonia was resolving. Patient also had a cough and was on occasions bringing up sputum.
- Gastrointestinal System: relatively normal. Patient had a NG tube.
- Genitourinary System: urine output was normal and not painful.
- Central Nervous System: normal.
- Musculoskeletal System: very weak due to the cachexia. Patient was unable to stand by himself.
- Endocrine System: normal.
- Others: pupillary light reflex and fundoscopy were normal.

Current therapy:

After the period in ITU and the occurrence of the blindness, the patient recovered considerably. However he was still given the antivirals for complete elimination of the CMV infection.

Discussion of results of general and specific examination:

The pupillary light reflex of the patient was normal, suggesting that his anterior visual pathway was intact and functioning normally. This indicated that the blindness was arising from a problem in the posterior visual pathway or maybe the occipital lobe. Patient was clearly blind however he insisted that he was seeing things normally. After some days he began to recognise some shades of color but other than that he remained blind.

General examination was normal. Chest was clear suggesting that the patient was recovering from the pneumonia.

Differential diagnosis:

- Regarding the pneumonia, at first it was thought that it was bacterial in nature, however diagnosis changed to CMV infection since CMV was detected by PCR.
- The blindness was probably caused by an injury to the brain during the patient's stay at ITU. Due to the complexity of the symptoms and their relative rarity, the diagnosis was still unknown.

Diagnostic procedures:

Laboratory Exams:

Test: Cell pathology.
Justification: Exclusion of malignancies.
Result: Scattered bronchial epithelial cells, many of which exhibited hyperplastic changes and few macrophages are present on a background of erythrocytes.
Conclusion: No malignant cells seen.

Instrumental Exams:

Test: CT Thorax.
Justification: Detection of pneumonia.
Results: Inflammatory changes of the lung are noted bilaterally basally and in the left upper lobe. Some pleural effusion is noted but there is no abscess formation. Lymphadenopathy is seen but there was no evidence of malignancy.
Conclusion: Pneumonia is confirmed.

Test: CT Brain.
Justification: Finding the cause of blindness.
Result: Multiple hypo dense zones of the brain are noted bilaterally.
Conclusion: These changes are most probably due to a post-infarctive process.

Test: MRI Head.
Justification: Finding the cause of blindness.
Result: Multiple bilateral abnormal cerebral foci. These appearances are suggestive of multifocal haemorrhages infarcts with associated haemorrhages changes, worse in the occipital regions bilaterally. Normal appearance of the Circle of Willis. No evidence of aneurysm and no evidence of venous sinus thrombosis.
Conclusion: Multifocal bilateral haemorrhagic infarcts.

Therapy:

Drugs:

Drug Name	Dosage	Frequency	Type	Reason
Lactulose	10mls	Dly	Synthetic, non-digestible sugar	Treatment of constipation
Omeprazole	40mg	Dly	PPI	Upset stomach

Patient received the last dose of oral anti-viral on the day of discharge.

Diagnosis:

Due to the pupillary light reflex being normal, it was concluded that the problem did not arise from the anterior visual pathway, as most cases of blindness do. The CT scans and MRI showed infarcts in the occipital lobe and this suggested that the problem was arising from the occipital cortex itself. Cortical blindness is defined as visual inability in the presence of normal light reflexes and normal fundoscopy[1]. This observation led to the diagnosis of Anton's syndrome, which is a very rare case of blindness. This syndrome is characterised by a phenomenon called confabulation[1]. Patients suffering from Anton's syndrome adamantly say that they can see when they obviously cannot and also speak as if they had normal vision[2]. This denial of vision problems is called visual anosognosia[2].

Cortical blindness was first described in detail by Gabriel Anton, who was an Austrian neurologist. This is why this syndrome now bears his name[3].

Final Treatment and Follow ups:

The treatment in this case is very limited. Follow-ups included visits from speech language pathologists, physiotherapy teams and occupational therapists. The patient was suggested to have a repeat CT Thorax, two weeks after the first one and to be regularly followed up at the stroke clinic.

Fact Box 5:

Title: Anton's Syndrome

Anton's Syndrome is a very rare type of brain damage that occurs in the occipital lobe. People with the condition suffer from cortical blindness.

Risk Factors: Since Anton's Syndrome is technically a stroke, its risk factors include:

- Hypertension
- Diabetes
- Smoking
- Alcohol consumption
- Prolonged periods of inactivity

Symptoms:

- Blindness (although the patient denies his blindness)
- Headaches
- Confusion.

Signs: A very specific sign of Anton's Syndrome is confabulation. Confabulation is a way by which the patient denies his blindness, walking and trying to move as he still sees normally. Confabulation is made very clear when the patient stars to describe objects and people in his vicinity that are in fact not there.

Prevention: This consists mainly in preventing cerebrovascular disease which include a healthy life style, controlled blood pressure, no smoking and alcohol consumption and others.

References:

1. Rickards C and Shepherd DI. Cortical blindness in a 35-year-old man. Postgrad Med J. 1996; 72(846): 249–251.
2. Maddula M, Lutton S and Keegan B. Anton's syndrome due to cerebrovascular disease: a case report. Journal of Medical Case Reports. 2009; 3: 9028.
3. Kondziella D and Frahm-Falkenberg S. Anton's Syndrome and Eugenics. J Clin Neurol, 2011; 7: 96-98.