Ophthalmic complications with disseminated intravascular coagulation

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SUMMARY Massive lid oedema, ecchymosis, proptosis with a total restriction of extraocular movement, markedly raised intraocular pressure, and occlusion of the central retinal artery developed acutely in the right eye of a 26-year-old woman with a past history of disseminated intravascular coagulation. She had been admitted to hospital for symptoms of abdominal pain and bleeding from multiple sites a few hours earlier. Five days previously she had some proptosis of the other eye and had been treated with antibiotics for suspected orbital cellulitis at another hospital. The oedema and proptosis resolved on high-dose intravenous corticosteroid therapy. Despite attempts to relieve the orbital oedema and raised intraocular pressure with a lateral canthotomy and antiglaucoma medications, the patient lost all perception of light in the right eye and has subsequently developed an optic nerve atrophy.

Disseminated intravascular coagulation (DIC) is a syndrome involving widespread intravascular coagulation, primarily in small vessels. Associated disorders have included head trauma, sepsis, surgery, burns, obstetrical complications, liver disease, malignancy, and inflammatory bowel disease. Common to these is the incitement of the coagulation cascade, with resultant thrombosis. Consumption of clotting elements eventually leads to hypocoagulability and haemorrhage. Reported ocular complications include thrombosis of choroidal and retinal vessels, as well as bleeding in the optic nerve sheath, vitreous, choroid, and retina. Similar problems are seen in the brain, heart, kidney, gastrointestinal and genitourinary tracts, as well as involvement of other organs.

Case report

A 26-year-old woman presented to the University Hospital with severe abdominal pain, headache, and bleeding from the urinary tract, rectum, vagina and nose. She had ecchymosis of the knees and buttocks and petechiae of the hard palate. Generalised oedema, most marked in the head and neck regions, and massive lid oedema with proptosis were present. Five days prior to this admission the patient had noted fever, proptosis, and lid oedema of her left eye and was admitted to another hospital. Although there was no evidence of sinusitis or a history of penetrating injury, she was treated with oral and intravenous antibiotics for orbital cellulitis. Orbital symptoms and signs progressed slowly despite treatment. On the day of transfer to the University Hospital the unaffected right eye became acutely proptotic.

She had a history of colitis and DIC. The first episode of DIC occurred two and one-half years previously during an active period of colitis when the patient was five months pregnant. Generalised oedema, including facial swelling, was present at that time. The patient improved on prednisone and sulphasalazine therapy. Six months before the present admission she had been in hospital again with DIC. She improved with the addition of prednisone to the maintenance sulphasalazine therapy. Ophthalmic complications were not present during either episode.

Our examination revealed a blind right eye, and uncorrected near vision of 20/200 in the left eye. Pronounced lid oedema, proptosis, and haemorrhagic conjunctival chemosis were present (Fig. 1). Intraocular pressure by Schiötz tonometry was
greater than 80 mmHg in the right eye and 11 mmHg in the left eye. There was no extraocular motility of the right eye; there was approximately 50% of normal abduction and adduction of the left eye. Ophthalmoscopic examination revealed narrowed retinal arterioles and diffuse retinal oedema with a cherry red macula in the right eye; the fundus of the left eye appeared normal. There was no evidence of intraocular inflammation in either eye.

Laboratory data from the referring hospital included a platelet count of $160 \times 10^9/l$, a prothrombin time (PT) of 25 seconds, a partial thromboplastin time (PTT) of 100 seconds, a fibrinogen level of 0.29 g/l, and a fibrin split product (FSP) titre of 1:2560. Repeat laboratory studies at the time of admission showed the platelet count had decreased to $69 \times 10^9/l$; the erythrocyte sedimentation rate was 7 mm/h (Westergren). Serological studies for antinuclear antibody titre, rheumatoid factor, and complement levels gave normal results. A pregnancy test was negative. Blood and conjunctival cultures were negative. Computerised tomographic examination of the orbits showed bilateral proptosis, thickening of the muscle outlines, and increased radiodensity of the optic nerves (Fig. 2).

Initial treatment included intravenous administration of 125 mg methylprednisolone followed by intravenous dexamethasone 25 mg every 6 hours for four doses and then 20 mg every 6 hours for four doses. A right lateral canthotomy with division of the canthal tendon was performed. No significant blood was drained by this manoeuvre, though the tenseness of the orbital tissues was noticeably relieved. In the subsequent seven hours the intraocular pressure in the right eye fell to 29 mmHg. Vision has remained no light perception in the right eye but improved to 20/40 in the left eye over 12 hours and to 20/20 over several days. Orbital oedema and proptosis gradually resolved, but the right eye slowly developed optic atrophy.

**Discussion**

Among the multiple extraintestinal manifestations of inflammatory bowel disease is hypercoagulability resulting from abnormal coagulation factors and platelets.\(^4\) This may have precipitated the episodes of DIC in our patient. Talbot et al.\(^5\) observed DIC in three patients with inflammatory bowel disease, one of whom also had sepsis. Endo et al.\(^6\) described DIC in two patients with ulcerative colitis; however, in both patients the DIC followed a surgical procedure.

This patient’s coagulopathy probably involved the ophthalmic, maxillary, and episcleral vessels, resulting in ischaemia, haemorrhage, and massive oedema. These factors may have led to an increased episcleral venous pressure, to raised intraocular pressure, and eventually to central retinal artery occlusion. In carotid-cavernous fistula the arterialisation of all orbital vessels may lead to an increased intraocular pressure.\(^7\) Presumably this occurs secondary to a raised episcleral venous pressure. Alternatively, optic nerve ischaemia or thrombus formation in the central retinal artery may have contributed to visual loss.

The extraocular muscle dysfunction presumably resulted from massive oedema, though ischaemia to the muscles themselves is a possibility. No restriction
was present on forced duction testing. Periorbital oedema has been described in a patient with thrombotic thrombocytopenic purpura. We can only speculate as to whether the mechanisms may have been the same.

Many arterial and venous complications have been reported as sequelae of inflammatory bowel disease with or without DIC, including central retinal arterial occlusion. Proptosis and lid oedema have been reported in a patient with orbital pseudotumour and Crohn’s disease; we believe our patient’s course to be too rapid and the oedema too massive to be explained on such a basis. Since our patient did not show any ocular signs of inflammation, we do not believe the inflammation associated with Crohn’s disease (that is, uveitis or optic neuritis) was a likely contributing cause of visual loss.

Other possible explanations of our patient’s course might include vasculitis or infection, but there were no clinical or laboratory data to support a diagnosis of either. Fever is known to occur in patients with colitis.

The ophthalmic complications we report here are rare in DIC. To our knowledge the sudden development of massive orbital oedema, markedly raised IOP, and occlusion of the central retinal artery has not been reported previously. Since the administration of high doses of intravenous corticosteroids decreased the oedema rather quickly, it is reasonable to assume that this therapy, along with adequate decompression undertaken shortly after the initial development of the orbital oedema, might have averted the subsequent series of ocular complications.

References


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