

# Occlusion of the retinal artery in two cases of orbital granuloma

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Orbital granulomata are a cause of proptosis of one or both eyes. Sometimes these granulomata are found to consist of non-specific inflammatory tissue with prominent perivascular foci of necrosis and a high mononuclear cell component. They are then thought to belong to the same group of disorders as Wegener's granulomatosis and lethal midline granuloma. Indeed, some cases of apparently isolated orbital granuloma have gone on to present the full picture of these diseases (Churg and Strauss, 1951; Stratsmaa, 1957).

Two cases of orbital granuloma, one certainly belonging to this group, are described in which the complication of a retinal artery thrombosis occurred.

## Case 1

**A man aged 64**, admitted under the care of Dr. R. W. Ross Russell, had a 3-week history of frontal headache which was throbbing in character and worse on the right side. A few days after the onset of headache he noticed double vision and the right eyelid started to droop. A severe pain developed in and around the right eye and vision began to fail in that eye. Two weeks later a similar process started in the left eye. He smoked  $\frac{1}{2}$  oz. tobacco daily and past history was negative except for pulmonary tuberculosis in childhood. There was no history of thyroid disease.

### *Examination*

The right eye was severely proptosed, with marked chemosis. There was complete ophthalmoplegia on the right and a firm bean-shaped mass could be palpated beneath the globe. The proptosis was not reducible by pressure on the globe, and orbital resistance was increased. Visual acuity was reduced to perception of light and the fundus showed slight swelling and scattered haemorrhages. The pupil was moderately dilated and fixed. The left eye was slightly proptosed and showed complete paralysis of abduction and marked limitation of vertical movements and adduction; pupil reactions were normal and the visual acuity 6/6 corrected. The left fundus was normal. The rest of the examination was negative except for an extensor plantar response on the right side. The corneal response was present on both sides. The patient was afebrile.

### *Investigations*

White cell count was normal, but the erythrocyte sedimentation rate was raised at 57 mm. in the 1st hour. The urine was normal and free from protein. Skull *x* rays revealed a craniovertebral junction deformity with prominent odontoid peg and an unfused and poorly-developed posterior

arch to the atlas vertebra. This was thought to explain the right extensor response. Views of the orbits were normal and so were bilateral carotid angiograms.

All other investigations were not helpful, and it was decided to biopsy the bean-shaped mass. This showed on histological examination (Prof. Blackwood) mainly collagenous tissue with many perivascular foci of mononuclear cells with eosinophils, plasma cells, and small areas of necrosis. This picture is morphologically similar to that of Wegener's granulomatosis.

#### *Treatment*

The patient was started on dexamethasone at a dose of 12 mg. daily. The day after starting treatment he complained of complete loss of vision in the right eye and on examination the fundus showed the picture of complete retinal artery thrombosis with stasis of the blood in the veins, a pale disc and very attenuated arteries (Figure). After this catastrophe the patient improved rapidly and after 5 days on dexamethasone the left eye was back to normal. In addition, the right-sided proptosis receded rapidly and some movement returned to the eye.



FIGURE Case 1 *Right fundus 3 hours after retinal artery occlusion*

After 2 weeks' treatment, the only signs remaining were a complete lateral rectus palsy on the right and of course total blindness in that eye. The erythrocyte sedimentation rate came down to 17 mm. in the 1st hour.

#### *Progress*

Dexamethasone was reduced gradually to 2 mg. daily. After one month the headache recurred and proptosis of the right eye followed once again. At the time of writing the patient has shown a second dramatic response to treatment with dexamethasone in high dosage, but has developed much muscle wasting and weakness involving both proximal and distal muscle groups of the limbs. Electromyographic studies have shown widespread involvement of the peripheral nerves, and a muscle biopsy has revealed a condition compatible with either quiescent polyarteritis or steroid-induced myopathy.

#### **Case 2**

**A man aged 65**, admitted under the care of Mr. Wylie McKissock, had noticed persistence of bilateral frontal headache after a febrile illness 2 months before admission; 3 days before admission the headache intensified and the left eye became very painful and swollen, with a marked deterioration of vision in that eye. The patient had worked in a steel factory some years previously, where he

had been in contact with sand dust. He smoked 15 cigarettes daily and suffered from chronic bronchitis.

#### *Examination*

There was a left-sided severe proptosis, not reducible by pressure, marked chemosis, and conjunctival injection. Eye movements were lost completely although the pupil still showed a consensual light reflex. No tumour was palpable in the orbit. Visual acuity was reduced to light perception on the left, and the fundus showed marked papilloedema with haemorrhages. The right eye was normal with a visual acuity of 6/6 corrected, unimpaired visual field, and full eye movements. An ear, nose, and throat examination was normal.

#### *Investigations*

The white cell count was normal, but the erythrocyte sedimentation rate was raised to 36 mm. in the 1st hour. X rays of the skull and orbits were normal, as was a left carotid angiogram. Sinus x rays showed opaque frontal, ethmoid, and sphenoid sinuses on the left side.

#### *Treatment*

Initially the patient was treated with high doses of chloramphenicol together with Cloxacillin and penicillin V. Soon after admission to hospital vision was totally lost in the left eye and the fundus was found to be very pale with marked attenuation of the arteries. Retinal artery occlusion was diagnosed.

At this stage he was started on dexamethasone at a dose of 16 mg. daily in addition to the antibiotics. As in the previous case there was a dramatic response and within 2 weeks the proptosis of the left eye completely receded. In this case there was no residual lateral rectus paralysis. The erythrocyte sedimentation rate fell to 18 mm. in the 1st hour. Dexamethasone was gradually reduced and eventually stopped altogether after 8 weeks.

#### *Progress*

At the time of writing 3 months after the original admission, there has been no recurrence.

### **Discussion**

Retinal artery thrombosis is not reported as complicating granulomata occurring from other causes; *e.g.* reparative granuloma (Sood, Malik, Gupta, and Kakar, 1967), foreign body granuloma (Wolter, Fralick, and Tanton, 1966), lipid granuloma (Eifrig, 1968), juvenile xanthogranuloma (Gaynes and Cohen, 1967), and giant cell granuloma (Cook, 1965). In the granulomata belonging to the Wegener's group of disorders, it is noted that foci of necrosis are perivascular in situation, and it is possible that the fundamental lesion is in fact an arteritis, which would predispose to the development of thrombosis. In support of this, cases of granuloma of the orbit have been reported (Harcourt, 1964; Hope-Robertson, 1956) in which there was an associated widespread necrotizing angiitis typical of polyarteritis nodosa. This spectrum of diseases, ranging from a purely local granuloma to a widespread necrotising angiitis, may therefore represent differing responses to the same basic lesions.

The important features of the two cases reported here were the rapid onset, the dramatic response to steroids, and the comparatively early occurrence of retinal artery thrombosis. Both patients experienced a great amount of pain in the affected orbit, a feature which also responded to steroids. The differential diagnosis of these cases included orbital cellulitis, orbital tumours, and, in the bilateral case, cavernous sinus thrombosis. The palpation of an orbital mass was an important finding in the first case and it allowed a firm diagnosis to

be reached by means of a biopsy. Both showed a raised erythrocyte sedimentation rate, but there were few other clues to the diagnosis. X rays, including special views of the orbit and contrast studies of the intracranial circulation, were not helpful. In the second case the ethmoid and sphenoid sinuses on the left were opaque and it may be that the granulomatous process had spread to these structures, although ear, nose, and throat examination of the nasopharynx was normal.

The response to steroid treatment was immediate in both cases and it is possible that the sight of the affected eyes of these two patients would have been saved had treatment been started earlier. It would seem reasonable to give a therapeutic trial of steroids in high dosage to cases of this kind, even if definite histological diagnosis cannot be reached. Antibiotic cover during steroid treatment is to be recommended in these cases, particularly where there is conjunctival infection and the diagnosis is not fully established.

### Summary

Two cases of granuloma of the orbit, one bilateral and one unilateral, are described. Both showed a rapidly developing painful proptosis and external ophthalmoplegia, with evidence of venous and lymphatic blockage. In one case the histology of the lesion was examined and found to resemble that of Wegener's granuloma. In both cases there was a rapid response to treatment with steroids, but both patients developed a central retinal artery thrombosis early in the course of the illness. The importance of early treatment of such cases with steroids is underlined.

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