Acute endocrine exophthalmos

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The onset of endocrine exophthalmos is generally slow and any variations are gradual. However, it can develop suddenly (within minutes—Dock, 1915; overnight—Falta, 1913). When assessing the progress of exophthalmos, it is important to use an exophthalmometer, as the degree of chemosis and the variations in the width of the palpebral fissure can be deceptive. Many reports of dramatic changes in the degree of exophthalmos lack the support of exophthalmometric readings.

It has been noted (Werner, 1966; Pinchera, Pinchera, and Stanbury, 1965; Liddle, Heysell, McKenzie, 1965; Eversman, Skillern, and Senhauser, 1966; Fox and Schwartz, 1967) that endocrine exophthalmos may occur in the euthyroid state and in patients who have never had thyrotoxicosis.

Case report

A 60-year-old man presented at the ophthalmic unit as an emergency on 11 March 1975. He had a 24-hour history of pain, redness, watering, swelling, and some deterioration of visual acuity in the right eye. He also complained of double vision. There was no history of trauma.

Ocular examination revealed a moderate degree of right axial proptosis. The visual acuity was 6/9. The conjunctiva was chemotic. No pulsation of the globe could be detected and the proptosis was virtually irreducible. There was no cranial bruit. Ocular movements of the right eye were grossly limited in all directions with almost complete absence of elevation. The cornea was intact and lid closure was full. Visual acuity in the left eye was 6/6 and this eye appeared to be normal. Exophthalmometer readings were 22 mm right eye, 17 mm left. Visual field examination showed no field defect (Goldmann perimeter). Skull $\times$ ray revealed a soft-tissue mass in the right orbit. The carotid pulses and fundi were normal.

General examination showed the patient to be rather sluggish, both mentally and physically, and he did not give a clear history of his illnesses. He did not seem to be unduly concerned about his eye condition. He had no fever and the pulse rate and blood pressure were normal.

A routine urine analysis showed a level of 2 per cent sugar and a random blood sugar was found to be 258 mg/100 ml. The patient then stated that he was a diabetic but 'had got over all that now'. His medical records showed that he had been a known diabetic for 4 years, controlled on 1000 calorie diet and one tablet of phenformin 50 mg daily. He had been a persistent defaulter from the diabetic clinic.

The patient was admitted to the eye unit. In view of the findings, the possibility of a localized orbital cellulitis was considered, and treatment was started with local and systemic antibiotics.

12 MARCH 1975

The proptosis in the right eye had increased, and, during the night, the left eye had also become proptosed, to an even greater extent than the right (Figs 1a, b, c). There was now chemosis on both sides and a marked restriction of ocular movements. The corneae remained intact, but lid closure was becoming difficult. Exophthalmometer readings were 24 mm right eye, 25 mm left. The intraocular pressure by applanation tonometry was 40 mmHg right eye, 28 mmHg left. Acetazolamide tablets 250 mg four times daily were added to the treatment. Because of the clinical picture now presenting, a diagnosis of acute endocrine exophthalmos was made.

13 MARCH 1975

The chemosis was more marked and the patient had great difficulty in closing his eyes. The corneae were intact (Figs 2a, b, c).

Treatment was started with systemic steroids, prednisolone 60 mg daily. Exophthalmometer readings were 24 mm right eye, 25 mm left. The visual acuity was 6/9 right eye, 6/12 left. Intraocular pressures were 36 mmHg right eye, 48 mmHg left.

14 MARCH 1975

The exophthalmos had further increased. The degree of chemosis was unchanged and the patient was still able to close both eyes, but with extreme difficulty. Both corneae remained intact. Drops of adrenaline 1 per cent twice daily were instilled into each eye. The right visual field showed slight constriction and a fundus examination revealed early papilloedema on both sides.

Exophthalmometer readings were 26 mm right eye, 27 mm left. The visual acuity was 6/9 right eye, 6/12

(1a)

(1b)

(1c)

FIG. 1 (a) *Frontal view on 12 March.* (b) *Right lateral view on 12 March.* (c) *Left lateral view on 12 March*

left. Intraocular pressures were 23 mmHg right eye, 32 mmHg left.

In view of the deterioration, it was felt that a decompression procedure might be necessary, and a consultation was arranged with a neurosurgeon for the next day. The dosage of prednisolone was increased to 120 mg daily (Kinsell, Partridge, and Foreman, 1953; Werner, 1966; Wright, 1970).

15 MARCH 1975

A marked improvement was noted. The chemosis was much less, the ocular movements were less restricted, and the visual fields had returned to normal. The papilloedema noted on the previous day had disappeared.

Treatment was continued with 120 mg prednisolone daily.

Exophthalmometer readings were 21 mm right eye, 24 mm left. The visual acuity was 6/9 right eye, 6/9 left. Intraocular pressures were 22 mmHg right eye, 22 mmHg left.

16 MARCH 1975

Improvement continued. The chemosis had nearly disappeared and the ocular movements were almost normal with a slight restriction of elevation.

Exophthalmometer readings were 21 mm right eye, 24 mm left. The visual acuity was 6/6 partial right eye, 6/6 partial left. Intraocular pressures were 20 mmHg right eye, 21 mmHg left.

FIG. 2 (a) Frontal view on 13 March. (b) Right lateral view on 13 March. (c) Left lateral view on 13 March

(2a)

(2b)

(2c)

17 MARCH 1975

Exophthalmometer readings were 20 mm right eye, 23 mm left. The visual acuity was 6/6 partial right eye, 6/6 partial left. Intraocular pressures were 17 mmHg right eye, 20 mmHg left. The dosage of prednisolone was reduced to 60 mg daily.

19 MARCH 1975

Exophthalmometer readings were 18 mm right eye, 23 mm left. The dosage of prednisolone was reduced to 30 mg daily.

21 MARCH 1975

Exophthalmometer readings were 18 mm right eye, 22 mm left.

24 MARCH 1975

Exophthalmometer readings were 18 mm right eye, 21 mm left.

25 MARCH 1975

The patient was discharged. He was prescribed prednisolone 20 mg daily, chlorpropamide 250 mg daily, and phenformin SR 50 mg twice daily. He was restricted to a 1000 calorie diet.

4 APRIL 1975

Exophthalmometer readings were 16 mm right eye, 17 mm left. The dosage of prednisolone was reduced to 15 mg daily.

(3a)

(3b)

(3c)

FIG. 3 (a) Frontal view on 22 April.
(b) Right lateral view on 22 April.
(c) Left lateral view on 22 April

22 APRIL 1975

The patient was reviewed at the ophthalmic unit. The chemosis had completely disappeared and the ocular movements were full (Figs 3a, b, c). Exophthalmometer readings were 15 mm right eye, 16 mm left. The visual acuity was 6/6 partial right eye, 6/6 left. The intraocular pressures were 16 mmHg right eye, 17 mmHg left. The dosage of prednisolone was reduced to 5 mg daily.

30 MAY 1975

Prednisolone was discontinued and the eyes remained normal. Exophthalmometer readings were 15 mm right eye, 16 mm left. The response is summarized in Fig. 4.

INVESTIGATIONS

Haemoglobin	16.7 g/100 ml
White blood count	10 600/cm
Blood urea	40 mg/100 ml

Sodium	131 mmol/l
Chloride	92 mmol/l
Potassium	3.6 mmol/l
Creatinine	0.7 mg/100 ml
CO ₂	21 mmol/l
Blood glucose	258 mg/100 ml
Bilirubin	1.5 mg/100 ml
Cholesterol	438 mg/100 ml
Plasma calcium	11.1 mg/100 ml
Uric acid	4.4 mg/100 ml
Alkaline phosphatase	87 mU/ml
Albumin	4.6 g/100 ml
Inorganic phosphate	4.1 mg/100 ml
Total protein	7.8 g/100 ml

The Wassermann reaction and Kahn test were negative. X rays of the right orbit, sinuses, and skull, were taken on 11 March and showed an opacity of soft tissue density in the lower part of the right maxillary antrum

FIG. 4 Graph showing ocular response to systemic steroids

with a smooth upper border. No associated bone destruction was identified and it seems likely that this was a polyp. There was a considerable difference in the density of the orbits, that on the right showing an overall opacity consistent with the soft tissue swelling. No definite bone destruction was seen. Thyroid function tests showed a thyopac 3 value of 118.8, and thyopac 4 of 11.8 μ g per cent, with a free thyopac index of 9.9 which is normal.

A thyrotrophin releasing hormone (TRH) test showed an impaired response which suggested partial thyroid autonomy. At 0 min thyroid stimulating hormone (TSH) was 1.6 mU/l, at 20 min 3.9 mU/l, and at 60 min 3.0 mU/l. Thyroid antibodies were all negative.

Discussion

The unusual features of this case were the rapidity of onset—the proptosis in the right eye developing probably within 24 hours, and that in the left within six hours—and the rapidity and completeness of the response to systemic steroids (Brown, Coburn, Wigod, Hiss, and Dowling, 1963).

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In other cases reported (Hoffenberg and Jackson, 1958; Brown and others, 1963; Werner, 1966) there was also rapid response to steroid therapy, but in only one case did the condition clear rapidly. Whether coincidentally or not, the patient was, like the case reported here, a clinically euthyroid diabetic. Further study into the possible connexion between acute endocrine exophthalmos and diabetes could prove fruitful.

Ideally, this condition should be treated by the ophthalmologist in collaboration with the physician; the former observing the state of the corneae, fundi, ocular fields, and the degree of exophthalmos, and the latter observing the patient's physical and biochemical response to the large doses of steroids.

It has been stated (Duke-Elder, 1974) that in the treatment of endocrine exophthalmos, large doses of systemic steroids usually need to be administered over a considerable period of time, with the inevitable hazard, not only of systemic complications, but also of the ocular side-effects such as cataracts and elevation of the intraocular pressure (Haddad, 1968).

Fortunately, in this case, the response was so rapid that it was possible to reduce the dosage before any such hazards were encountered.

Three months after complete cessation of drug therapy, there was no evidence of any recurrence of the condition. Whether this resolution will be permanent remains to be seen.

Summary

A case is reported of acute endocrine exophthalmos in a 60-year-old clinically euthyroid male diabetic and the rapid response to steroid therapy is described. The exophthalmometer was used to assess objectively the variations of the exophthalmos.

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