SUPERIOR ORBITAL FISSURE SYNDROME OF UNCERTAIN AETIOLOGY*

REPORT OF TEN CASES

BY

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DUKE-ELDER (1949) stated that "a considerable number, probably some 15 to 20 per cent. of cases of ophthalmoplegias always remain undiagnosed and must be classified as of uncertain aetiology despite the most careful investigations".

Roger and Alliez (1935) collected fifty cases of superior orbital fissure syndrome from the literature and added fifteen of their own; in 27 the cause was orbital apex neoplasm, seventeen were syphilitic, and many were of uncertain presumably inflammatory origin. Rucker (1958) gave statistics of 1,000 cases of ocular nerve paralysis, and showed that 3rd and 6th nerve affections were common. Paralysis of the 4th or combined 3rd, 6th, 4th nerves were infrequent. The causes were trauma to the head 17 per cent. ; cerebral neoplasms 17 per cent. ; vascular diseases and diabetes 15 per cent. ; aneurysms of the circle of Willis 11 per cent. ; infections, poliomyelitis, syphilis, and disseminated sclerosis 12 per cent., and undetermined in 28 per cent.

Unilateral ophthalmoplegias of uncertain origin may present in various ways:

(1) The superior orbital fissure syndrome, first described by Rochon-DuVigneaud (1896), is characterized by local pain, proptosis, paralysis of the 6th, 4th, 3rd, and first division of the 5th nerves, with neuralgic pains, and anaesthesia of the upper lid, side of the nose, forehead, temple, conjunctiva, and cornea.

(2) The partial superior orbital fissure syndrome may present as complete or partial 3rd or 6th nerve paralysis, usually accompanied by affection of the naso-ciliary nerve giving neuralgic pain and anaesthesia of the cornea and of the skin and conjunctiva of the medial canthus. Paralysis of the 4th nerve alone is rare.

(3) The orbital apex syndrome is a superior orbital fissure syndrome with involvement of the optic nerve in retrobulbar neuritis or papilloedema which may be followed by atrophy.

Collier (1921) described a sphenoidal fissure syndrome in patients at any age from puberty onwards which was due to transient periostitis of the sphenoid fissure. In many cases he attributed the condition to exposure to cold or to septic nasal sinuses; but in many others the cause was not obvious. He compared these cases to cases of common Bell's palsy. Almost all his

* Received for publication December 1, 1960.
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forty cases recovered completely in a period varying from a few weeks to a few months. In one case the optic nerve was involved. von Graefe (1866), Landesberg (1880), Ulthoff (1886), Suckling (1886), Wilson (1921), and others have described paralysis of the 6th nerve and complete ophthalmoplegia after exposure to cold or wet, which they attributed to acute infective (rheumatic) neuritis of the peripheral ocular nerves. Holmes (1931) suggested that the change of temperature lowered the resistance to a virus.

Jefferson (1947) found that of 158 intracranial aneurysms, 55 caused isolated paralysis of the oculomotor nerve with persistent unilateral pain in the distribution of the trigeminal, the 3rd nerve never regaining complete function. Hyland and Barnett (1954) studied 35 cases of oculomotor nerve paralysis caused by cerebral aneurysms; in eighteen the oculomotor nerve was compressed directly by the aneurysm, and in the remaining seventeen the nerve was involved by some secondary mechanism produced by haemorrhage. Henderson (1955) showed that angiography was positive in 94·5 per cent. of 91 patients; the aneurysm was missed in six patients, being obliterated by thrombosis.

Present Investigations

In the four cases of unilateral complete superior orbital fissure syndrome, three cases of incomplete superior orbital fissure syndrome, and three cases of orbital apex syndrome to be described in this paper, the general, ocular, rhinological, and neurological investigations were negative, and the cause of ophthalmoplegia remained uncertain.

Unilateral oculomotor nerve paralysis developed quickly with varying degree of headache, pain in the eye, and neuralgia of the forehead and temporal region on the affected side. The condition did not date from birth. There was no history of head trauma, strain, migraine, syphilis, or rheumatic pains. The patients were not alcoholics or taking drugs such as barbiturates to cause intoxication. There was no recent history of fevers such as diphtheria, influenza, measles, small pox, typhoid, typhus, mumps, malaria, whooping cough, septicaemia, cerebrospinal meningitis, poliomyelitis, or encephalitis, nor of a neurological disease or loss of consciousness.

The patients' general health was perfect. There were no signs of pellagra, beri-beri, or scurvy, and no allergic or hysterical manifestations. The skin of the face did not show any boils or herpetic eruptions. There was no enlargement of parotid, lacrimal, thyroid, or lymph glands, and no malignant tumour in the body. The temperature, blood pressure, pulse, and heart were normal. The chest did not reveal any tuberculous lesion. There were no foci of infection, such as septic teeth, etc. The tonsils and nasal sinuses were normal, with no sign of otitis media or nasopharyngeal tumours.

Apart from the unilateral ophthalmoplegia there was no other neurological manifestation. The motor and sensory systems, reflexes, and sphincters were normal. There was neither subjective or objective evidence of increased
intracranial pressure nor any endocrine or trophic changes. The clinical test for myasthenia gravis was negative and there was no lethargy. There was no sign of disseminated sclerosis after prolonged observation.

There was no oedema of the lids or chemosis of the conjunctiva to account for an acute orbital inflammation; and no subconjunctival haemorrhage or lid ecchymosis. Apart from cases of orbital apex syndrome showing papilloedema or optic atrophy on the affected side, there were no fundus or field changes. There was no arteriosclerosis of the retinal blood vessels, and no nystagmus or palpable orbital mass.

Urine was free of sugar, acetone, and albumin. The stools were negative for parasites. The blood Wassermann reaction was negative. The haemoglobin percentage, blood counts, erythrocyte sedimentation rate, blood sugar curve, bleeding and clotting time were normal. The Mantoux test was usually negative. The basal metabolic rate was normal. The cerebrospinal fluid, pressure, cells, protein, and salt were normal, and showed no red blood corpuscles.

A lateral x ray of the skull did not reveal changes in the sella turcica or clinoid processes, or any suprasellar or sellar calcification. The sphenoid sinus was normal. A postero-anterior view with 20° tube tilt showed no change denoting osteoperiostitis in the bony margin of the superior orbital fissure, or in the lesser or great wings of sphenoid. The anterior ethmoidal, maxillary, and frontal air sinuses were normal. Right and left oblique views showed normal optic canals and posterior ethmoidal air cells. A view of the base of the skull showed normal anterior and posterior ethmoidal and sphenoidal sinuses. The nasopharynx showed no abnormality. Lateral and postero-anterior angiograms did not show any sign of intracranial aneurysm.

The orbital apex was explored in most of the cases, to ascertain the cause. A lateral transconjunctival orbitotomy, after an external canthotomy (Knapp, 1874; Reese, 1941) was used. The intermuscular fascia between the lateral and inferior recti was cut, and the little finger palpated gently the apex of the orbital muscle space around the optic nerve, especially on its lateral side, to find the cause.

Case Reports

I.—Superior Orbital Fissure Syndrome

Case 1, a 32-year-old single female (Fig. 1, opposite), had a right superior orbital fissure syndrome of one week’s duration. The visual acuity was 6/6. There was right proptosis 21 mm. (left 16 mm.) The fundi were normal. Apart from paralysis of the right 3rd, 4th, 6th, and 1st branch of the 5th nerves there were no ocular or neurological findings. The history and investigations were not suggestive of the causal lesion. The case was treated with vitamin C and B complex, calcium, and iodides. After 4 months the condition began to improve, and proptosis and ptosis disappeared. The right pupil became normal in size and reactions, and the ocular movements to the left and to the right were
perfect. Upwards and downwards ocular movements were still defective, but these were regained in the course of a further 2 months.

**Case 2,** a 10-year-old male, had left partial ptosis and inability to move the left eye inwards, for 2 days. In another 2 days, while using antibiotics and vitamin C and B complex, the condition progressed to a complete superior orbital fissure syndrome. The left eye showed proptosis 25 mm. (and the right 16 mm.) and could not be moved upwards (Fig. 2), downwards, or sideways. The visual acuity was 6/6, and the fundi were normal.

There were no other neurological findings. The investigations did not point to any diagnosis. Angiography did not reveal any intracranial vascular abnormality. Orbital exploration of the apex of the muscle cone space revealed a mass on the postero-lateral side of the optic nerve extending to the apex of the orbit. It measured 1 x 1 cm., and was smooth, tense, and difficult to separate from the surrounding tissues by little finger dissection. On separating it with scissors, dark blood was evacuated and the mass immediately disappeared. A blood cyst under tension at the apex of muscle cone space was diagnosed. One week after the puncture of the cyst the proptosis and ptosis disappeared. The left pupil became normal in size and reactions, and the patient could move his left eye sideways, downwards, and upwards.

**Case 3,** an 8-year-old girl (Fig. 3), had had a left superior orbital fissure syndrome for one week. The history and investigations did not suggest any diagnosis. The visual acuity was 6/9, and the fundi were normal. Proptosis on the left side measured 25 mm. (and on the right 16 mm.).

Exploration of the orbital apex with the little finger revealed a smooth tense mass 1 x 1·5 cm. adherent to the postero-lateral side of the optic nerve. On separation of
the edges of the mass with scissors, altered blood came out, and the mass immediately disappeared. A blood cyst under tension at the orbital apex was diagnosed. After another week the patient recovered.

Case 4, a 36-year-old woman (Fig. 4), had had a left superior orbital fissure syndrome for 2 months. The history and investigations were negative. The visual acuity was 6/24, and with -2 D sph. 6/9. The fundi were normal.

The left proptosis measured 20 mm. (and the right 16 mm.). For 2 months the aetiology of the ophthalmoplegia was uncertain, and after 2 more weeks the proptosis increased and a hard mass was felt between the globe and the orbital roof. Surgical exploration of the orbit revealed a pink mass measuring 1 x 3 cm. extending backwards towards the superior orbital fissure. Histological examination showed a chronic non-specific inflammation associated with fibrosis. The inflammatory cells consist of lymphocytes and plasma cells, with a small number of eosinophils, mainly concentrated around blood vessels. The picture is consistent with a diagnosis of inflammatory pseudotumour.

II.—PARTIAL SUPERIOR ORBITAL FISSURE SYNDROME

Case 5, a 22-year-old male, complained of binocular diplopia on looking to the right, and severe headache and pain in the left eye for one week. Examination revealed left lateral rectus paralysis and anaesthesia in the distribution of the left nasociliary nerve, i.e. of the cornea, and skin and conjunctiva of the medial canthus, and limitation of inward movements of the left eye (Fig. 5). The pupillary reactions were intact, and ocular movements to the left were perfect. The visual acuity was 6/6, and the fundi were normal. The history and investigations were negative.

Using iodides, vitamin C, and calcium, the patient was cured in 3 weeks before orbital exploration was attempted.

Case 6, a 16-year-old girl (Fig. 6, opposite) had had a right 3rd nerve paralysis and anaesthesia in the distribution of the right nasociliary nerve for 2 weeks. The right eye was proptosed 21 mm. (and the left 17 mm.). The visual acuity was 6/6, and the fundi were normal. The history and investigations were negative. Right orbital exploration showed a tense blood cyst measuring 1 x 1 cm. lying beside the optic nerve at the apex of the muscle cone space. Puncture of the cyst evacuated altered dark blood and the patient was cured.
Superior orbital fissure syndrome

Case 6, a 16-year-old girl (Fig. 6) complained of diplopia on looking to the left, accompanied by severe neuralgic pain in left side of face and headache for 12 days. The visual acuity was 6/9, and the fundi were normal. There was left 6th nerve paralysis with limitation of laevoversion in the left eye. The other ocular movements were normal.

Case 7, a 30-year-old male (Fig. 7) complained of diplopia on looking to the left, accompanied by severe neuralgic pain in left side of face and headache for 12 days. The visual acuity was 6/9, and the fundi were normal. There was left 6th nerve paralysis. The other ocular movements were normal.

Case 7, a 30-year-old male (Fig. 7) complained of diplopia on looking to the left, accompanied by severe neuralgic pain in left side of face and headache for 12 days. The visual acuity was 6/9, and the fundi were normal. There was left 6th nerve paralysis with limitation of laevoversion in the left eye. The other ocular movements were normal.

There was also anaesthesia in the distribution of nasociliary nerve. The history and investigations were negative. A cure was effected in one month by the use of Diamox, iodides, and vitamin B complex.

III.—Orbital Apex Syndrome

Case 8, a 13-year-old girl (Fig. 8), complained of right proptosis that had begun with vomiting 2 months before, and had previously occurred twice at the ages of 6 and 10. On each occasion the proptosis had persisted for 3 months, and had then subsided. The nose was depressed after an injury in childhood. Examination showed a right orbital apex syndrome. The right proptosis measured 25 mm. (and the left 17 mm.). The right fundus showed post-papilloedemic optic atrophy. The visual acuity was counting fingers at 20 cm. The right pupil was dilated and inactive. There was no palpable orbital mass. The left fundus was normal, and the visual acuity 6/6.

Apart from the vomiting, the history and investigations were negative. Exploration of the right orbit revealed a tense cystic swelling measuring 1 x 1 cm. within the apex of the muscle cone space attached to the upper surface of the optic nerve. On blunt finger dissection the cyst ruptured and altered chocolate-coloured blood was evacuated that stained the orbit and subconjunctival tissues. One week later the proptosis and ptosis had disappeared, and the ocular movements of the right eye were normal. As this case showed, vomiting is a frequent symptom in acute orbital haemorrhage, the trigemino-vagus stimulation exciting an oculo-gastric reflex (Rollet and Paufique, 1929).
Case 9, a 20-year-old girl, complained of diminution of vision, proptosis, and ptosis in the right eye for 20 days. She noticed the proptosis first, and this was followed in the course of the day by ptosis and blurring of vision. Examination showed a right orbital apex syndrome with complete absence of ocular movement in the right eye (Fig. 9). The proptosis in the right eye measured 21 mm. (and in the left 16 mm.). The right fundus showed papilloedema, and the visual acuity was counting fingers at 50 cm.

The left fundus was normal, and the visual acuity 6/6.

The history and investigations were negative. Exploration of the right orbital apex revealed a tense cystic swelling measuring 1 x 1 cm. on the lateral surface of the optic nerve. Little finger dissection effected the evacuation of dark blood. After one week the proptosis and ptosis were cured, the ocular movements improved, the papilloedema disappeared, and the visual acuity was 6/12.

Case 10, a 55-year-old farmer, complaining of headache and severe neuralgic pain on the right side of the face, was admitted to the neurological section as a case of trigeminal neuralgia. The fundi were normal and there was no proptosis or ophthalmoplegia. The visual acuity was 6/9 in the right eye.

The left cornea was cloudy and the visual acuity 6/60.

3 months later the patient developed a right orbital apex syndrome (Fig. 10).

The right fundus showed optic atrophy, and the visual acuity was reduced to counting fingers at 50 cm., with a proptosis of 22 mm. (the left being 16 mm.). The history and investigations were negative, apart from a positive tuberculin test. Because of the right ophthalmoplegia and headache, a non-fistulous internal carotid aneurysm in the cavernous sinus was suspected, but this was ruled out by angiography, and the cause of the ophthalmoplegia remained uncertain for about 4 months. But later the proptosis increased and a hard nodular mass was felt between the globe and the upper orbital margin. Surgical exploration of the orbit showed an irregular pink non-encapsulated mass partly hard and partly soft, measuring 3 x 2 c. and extending around behind the optic nerve as far as the orbital apex. Histological examination showed tuberculous granulation tissue with a typical giant cell system and areas of caseation, and the diagnosis of tuberculoma of the orbit was confirmed.
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Discussion

As examples of ophthalmoplegia of unknown aetiology, four cases of superior orbital fissure syndrome, three cases of partial superior orbital fissure syndrome, and three cases of orbital apex syndrome have been described (Table). The orbital apex was explored in most of the cases to find the cause.

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Case No.</th>
<th>Age (yrs)</th>
<th>Sex</th>
<th>Side</th>
<th>Nerve Affected</th>
<th>Orbital Apex Lesion</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior Orbital Fissure Syndrome</td>
<td>1</td>
<td>32</td>
<td>Female</td>
<td>Right</td>
<td>Not explored</td>
<td>Cured</td>
<td></td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>10</td>
<td>Male</td>
<td>Left</td>
<td>Blood cyst (punctured)</td>
<td>Cured</td>
<td></td>
</tr>
<tr>
<td></td>
<td>3</td>
<td>8</td>
<td>Female</td>
<td>Left</td>
<td>Blood cyst (punctured)</td>
<td>Cured</td>
<td></td>
</tr>
<tr>
<td></td>
<td>4</td>
<td>36</td>
<td>Female</td>
<td>Left</td>
<td>Pseudotumour (removed)</td>
<td>Improved</td>
<td></td>
</tr>
<tr>
<td>Partial Superior Orbital Fissure Syndrome</td>
<td>5</td>
<td>22</td>
<td>Male</td>
<td>Left</td>
<td>Partial 3rd and nasociliary</td>
<td>Not explored</td>
<td>Cured</td>
</tr>
<tr>
<td></td>
<td>6</td>
<td>16</td>
<td>Female</td>
<td>Right</td>
<td>3rd and nasociliary</td>
<td>Blood cyst (punctured)</td>
<td>Cured</td>
</tr>
<tr>
<td></td>
<td>7</td>
<td>30</td>
<td>Male</td>
<td>Left</td>
<td>6th and nasociliary</td>
<td>Not explored</td>
<td>Cured</td>
</tr>
<tr>
<td>Orbital Apex Syndrome</td>
<td>8</td>
<td>13</td>
<td>Female</td>
<td>Right</td>
<td>Blood cyst (punctured)</td>
<td>Blood cyst (punctured)</td>
<td>Cured</td>
</tr>
<tr>
<td></td>
<td>9</td>
<td>20</td>
<td>Female</td>
<td>Right</td>
<td>Blood cyst (punctured)</td>
<td>Tuberculoma</td>
<td>Cured</td>
</tr>
<tr>
<td></td>
<td>10</td>
<td>55</td>
<td>Male</td>
<td>Right</td>
<td>Blood cyst (punctured)</td>
<td>Improved</td>
<td></td>
</tr>
</tbody>
</table>

Cases of superior orbital fissure and orbital apex syndromes of unknown aetiology are usually regressive and curable, and rarely accompanied by progressive proptosis.

Orbital apex exploration showed that in many cases the ophthalmoplegia was due to a small blood cyst under tension at the orbital apex pressing the ocular nerves against the superior orbital fissure edges. The origin of blood cyst is spontaneous haemorrhage from thin-walled capillaries, which may follow an unnoticed strain, similar to cases of spontaneous subconjunctival haemorrhage. The extent of the ophthalmoplegia varies from 3rd or 6th nerve paralysis usually accompanied by affection of the nasociliary nerve, to the complete superior orbital fissure syndrome or orbital apex syndrome according to the site and severity of the pressure.
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Stallard (1958) noted that pressure from blood after trauma within the superior orbital fissure might cause ophthalmoplegia, and that absorption of the blood resulted in recovery. Orbital blood cysts with absence of the endothelial lining resulting from a breakdown of a haematoma have been reported by Denig (1902), d'Amico (1924), Wheeler (1937), Svoboda (1948), and other authors.

The orbital muscle cone space is limited anteriorly by the union of the intermuscular membranes with Tenon's capsule. Charpy and Clermont (1911) showed that effusions in this space did not involve the lids or conjunctiva, the only leakage being towards the apex of the orbit. Blood in this space does not show as subconjunctival haemorrhage or ecchymosis of the lids, but leaks towards the superior orbital fissure. The encysted blood may then press on the ocular nerves causing ophthalmoplegia and even optic nerve lesions.

Many cases of recoverable ophthalmoplegia were attributed to sphenoidal or ethmoidal sinusitis, causing reactionary orbital oedema, toxic neuritis, or an affection of the superior orbital fissure. These lesions may result in isolated or multiple nerve paralysis which may recover (Dimsdale, 1948; Phillips, 1948) or in the orbital apex syndrome (Trantas, 1893; James, Thomson, Colledge, and Hodgson, 1936; Kjoer, 1945). Dimsdale and Phillips (1950) reported four cases of ocular palsy and six cases of superior orbital fissure syndrome which recovered spontaneously and were due to sphenoidal and posterior ethmoidal sinusitis. Perhaps in some of these cases, exploration of the apex of the orbit would have shown the presence of encysted blood at the apex of the muscle cone space pressing the ocular nerves.

Ophthalmoplegia, with progressive proptosis of uncertain origin, as in Cases 4 and 10, may be due to tuberculoma, inflammatory pseudotumour, or tumour beginning at the orbital apex. In the course of time the orbital mass usually extends forwards and becomes palpable, and biopsy then reveals the cause of the ophthalmoplegia.

Treatment

Unilateral partial or complete superior orbital fissure syndrome and orbital apex syndrome of uncertain aetiology may be treated medically by iodides, calcium, vitamins C and B complex, and Diamox. Many of these cases are due to encysted blood pressing the ocular nerves in the region of superior orbital fissure. This blood is usually absorbed in 3 weeks to 4 months according to the severity and rate of its absorption, and the ophthalmoplegia is then cured. Incomplete recovery may be due to nerve damage or fibrosis. In cases of incomplete superior orbital fissure syndrome, especially when the eye is not proptosed, digital exploration of the orbital apex through a lateral canthotomy is usually impossible, owing to the diffi-
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culty of introducing the little finger between the lateral orbital wall and the
globe. Kronlein’s operation is unnecessary as these cases usually recover.
Exploration of the orbital apex is indicated in the following cases:
(1) Orbital apex syndrome, before optic atrophy occurs.
(2) Complete superior orbital fissure syndrome if medical treatment is un-
successful after about 4 months, for fear of permanent damage to the ocular nerves.
In these cases a blood cyst is usually found at the apex of the muscle cone space,
and puncture of the cyst usually results in recovery.
(3) Ophthalmoplegia with progressive proptosis, especially when a palpable
orbital mass is present.

Summary
Cases of partial or complete superior orbital fissure syndrome and orbital
apex syndrome of unknown aetiology are usually regressive and curable.
They are rarely accompanied by progressive proptosis.
(1) Exploration of the apex of the orbit showed that many of these ophthal-
mopegias are due to spontaneous haemorrhage giving rise to encysted blood
under tension which presses the ocular nerves against the superior orbital
fissure edges. The extent of ophthalmoplegia varies with the site and
severity of the pressure. Absorption of blood in 3 weeks to 4 months is
usually followed by recovery.
(2) Ophthalmoplegia of uncertain origin with progressive proptosis may be
due to tuberculoma, inflammatory pseudotumour, or a tumour beginning
at the orbital apex.

REFERENCES

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doi: 10.1136/bjo.45.10.662

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