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Communications

Venous malformations in the orbit

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The purpose of this paper is to present the clinical, radiological, and pathological findings in twelve patients presenting with orbital varices and to draw attention to the syndrome of congenital venous malformation in the orbit: a condition which may be associated with venous malformations elsewhere in the body. With the introduction of modern radiological methods of contrast investigation, particularly orbital venography, a more detailed demonstration of these lesions in the orbit and their vascular connections is now possible.

Case material

Nine patients seen in the *X* ray Department at the City Road Branch of Moorfields during the period of 1966–1970 have been shown to have venous malformation in the orbit, either by radiology or at surgical exploration. All these were investigated by angiography—either by carotid angiography, or by orbital venography, or both. Three other cases seen before this period and described by Lloyd (1965) have been added to the series; they did not have angiographic studies but are included because of their clinical and radiological similarity to the other nine cases.

The clinical, radiological, and pathological details are set out in the Table (overleaf), and may be summarized as follows:

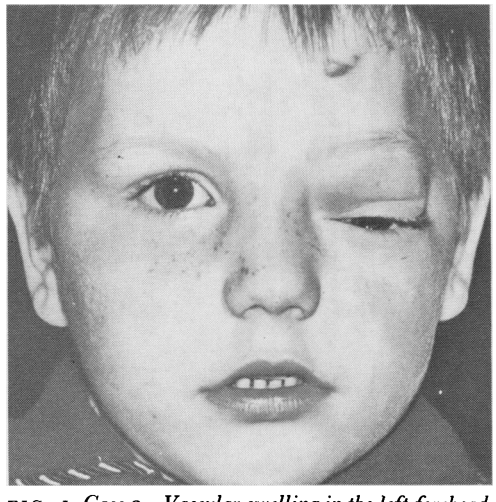

FIG. 1 *Case* 3. *Vascular swelling in the left forehead*

Clinical findings

The characteristic clinical picture was that of a child, adolescent, or young adult with a history of proptosis usually dating from birth or early childhood. The proptosis was normally provoked or made worse by an increase in the venous pressure in the head and neck. Vascular abnormalities were present in 75 per cent. affecting the conjunctiva or eyelids, and in three patients abnormal veins and localized venous dilatations were found on the forehead and scalp (Fig. 1). In one patient (Case 4) there was a venous abnormality of the hard palate.

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Table

Case no.	Age (yrs)	Sex	Clinical summary	X ray findings		Operations	Pathology
				Plain films	Contrast studies		
1	29	M	Proptosis and occasional diplopia since childhood Soft reducible mass present in inner part of upper eyelid.	Enlarged orbit with solitary orbital phlebolith Abnormal venous 'lakes' in frontal bone		Orbital decompression only No full exploration of the orbit	
2	15	F	Protrusion and redness of the right eye from birth Gradual increase in severity of signs despite radiotherapy Fullness of right upper lid with enlarged venous malformation at inner canthus Partial iris coloboma with episcleral vascular swelling in lower fornix extending to limbus from 3 to 6 o'clock Right optic atrophy	Enlarged orbit with solitary extraorbital phlebolith in pterygoid plexus Abnormal venous 'lakes' in frontal bone		Excision of conjunctival and episcleral varices Catholysis to enlarged veins in upper lid	Lesion composed of dilated venous channels lined by single layer of endothelium Few arterioles Prominent connective tissue stroma with lymphocytes
3	5	M	Vascular swelling of left forehead, both eyelids, and left orbit from birth 5 mm. proptosis Left globe displaced downwards 6 mm., laterally 4 mm. (Fig. 1)	Orbit enlarged Abnormal venous 'lakes' in frontal bone	Carotid angiography showed multiple intracranial arteriovenous malformations No abnormality in affected orbit apart from abnormal origin of middle meningeal artery Orbital venography (at orbitotomy) showed large varix (Fig. 2)	Extended lateral orbitotomy revealed grossly enlarged superior orbital venous system with extension of enlarged vessels round levator and superior rectus muscles. Abnormal veins excised and main vein ligated at orbital apex	Lesion composed of dilated venous channels and many small arteries (Fig. 3)
4	13	M	Red right eye from birth Fullness of right upper eyelid with subconjunctival vascular swelling near nasal and temporal limbus Right eye was proptosed 3 mm., displaced downwards 4 mm., laterally 3 mm. Associated vascular abnormalities on right side of hard palate and scalp	Enlarged orbit with multiple phleboliths in orbit (Fig. 4) Abnormal venous 'lakes' in frontal bone	Arteriography showed displacement of ophthalmic artery only Injection of superficial varix showed abnormal collection of venous channels in orbit (Fig. 5)	No surgery	
5	11	M	Swelling of right lower lid noticed at age 3 Bluish swelling deep to skin of right lower lid enlarged on Valsalva No proptosis	Soft tissue swelling visible with solitary phlebolith	Venography showed small varix demonstrated by direct injection (Fig. 6)	Varix was excised through incision in inferior fornix	Lesion consists of dilated and thrombosed vein

			looking up Right eye proptosed 3 mm. and displaced downwards 2 mm. Signs and symptoms gradually resolved over 3 wks Two previous similar episodes at age 14 and 17	Abnormal vascular calcification	' vascular calcification Venography failed		
7	16	F	Increasing proptosis for 10 years with occasional sudden increases in degree of proptosis Subconjunctival vascular malformations Right eye displaced downwards 3 mm.	Enlarged orbit with two phleboliths	Arteriography failed to show any abnormality Venography showed dilated superior ophthalmic vein and partial filling of varix medially in orbit (Figs 7 and 8)	No surgery	
8	26	F	Left proptosis since birth with subconjunctival vascular swellings	Enlargement of left orbit	Arteriography not performed Venography showed abnormal veins in lateral and posterior part of orbit (Fig. 9)	No surgery	
9	74	F	Enlarged left upper eyelid since childhood Recent deterioration of vision and heaviness of eyelids Swelling of left upper eyelid enlarged on stooping and Valsalva Subconjunctival varices in left upper and lower fornices	Enlargement of left orbit with numerous phleboliths Abnormal calcification immediately above and to left of pituitary fossa		No surgery	
10	54	M	Periodic redness and watering of right eye for 20 years Right-sided proptosis of 4 mm.	Negative	Orbital venography showed a collection of varices in line of inferior ophthalmic vein communicating with pterygoid plexus (Fig. 10)	No surgery	
11	20	M	Double vision on looking left for 4 yrs Right eye proptosed 5 mm. and displaced downwards 3 mm. Elevation of globe restricted	Negative	Arteriography showed varix in anterior part of orbit and reflux filling of superior ophthalmic vein from cavernous sinus on later films Orbital venography showed no filling of orbital veins on affected side	Lateral orbitotomy showed an abnormally distended venous system in posterior part of orbit	Venous anomaly composed of thickened and dilated varicose channels one thrombosed One channel occluded by compression due to stromal fibrosis prominent throughout lesion Vein walls composed predominantly of connective tissue with sparse elastic fibres Thin internal elastic lamina well seen (Figs 11, 12, 13)
12	17	F	1 mth swelling in left upper lid Similar swelling noticed twice before, each lasting 2 to 3 mths Cystic firm blue subcutaneous swelling near trochlea No proptosis or displacement of globe	Negative	Non-filling of first part of superior ophthalmic vein	Thrombosed varix of first part of left superior orbital vein excised through brow incision	Simple dilated venous channel containing thrombus (Fig. 14)

Radiological findings

PLAIN X-RAY CHANGES

These varied with the extent of the venous abnormality, but the following signs were apparent in this series.

(1) Enlargement of the affected orbit (8 patients).

(2) Presence of one or more phleboliths in the orbit (6 patients) or extraorbital phleboliths (1 patient).

(3) Large venous lakes or vascular markings in the frontal bone on the affected side corresponding to the venous enlargements found clinically (4 patients).

(4) In one case there was vascular calcification in the orbit (Case 6) and in another (Case 9) a similar type of calcification was observed intracranially above the pituitary fossa.

CONTRAST INVESTIGATION

On carotid angiography no arterial abnormality was found other than displacement of vessels in the orbit, but in two patients it was possible to demonstrate abnormal veins in the orbit by subtraction studies of the later venous phase of the arteriogram (Cases 6 and 11). As might be expected the most diagnostically rewarding examination was orbital veno-

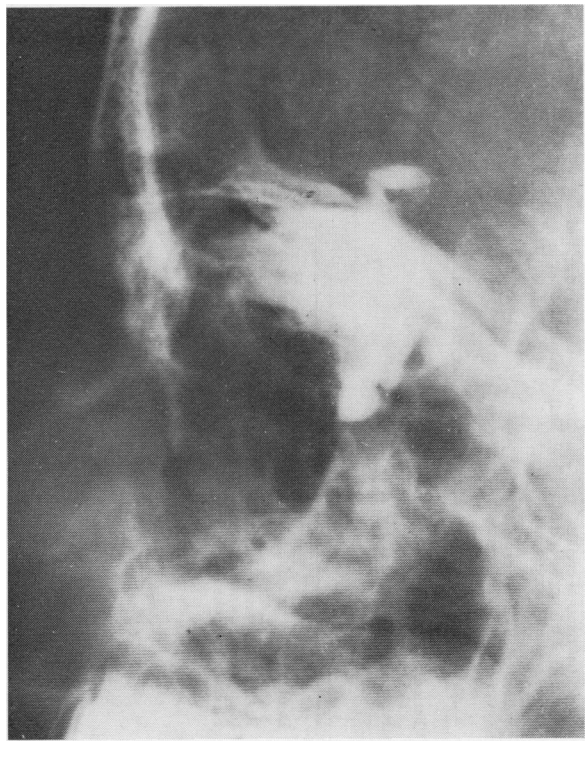

FIG. 2 *Case 3. Lateral radiograph of orbit, showing a large varix; contrast medium was injected directly into varix at orbitotomy*

graphy, and eight patients were examined by this method. In some only a solitary varix (Fig. 2) was demonstrated, while in others there was a collection of abnormal venous spaces in the orbit. (Fig. 5). The varices had no direct connection with the arterial side of the circulation and were often, to a large extent, divorced from the main venous circulation in the orbit. The presence of phleboliths implies venous stagnation (Hager,

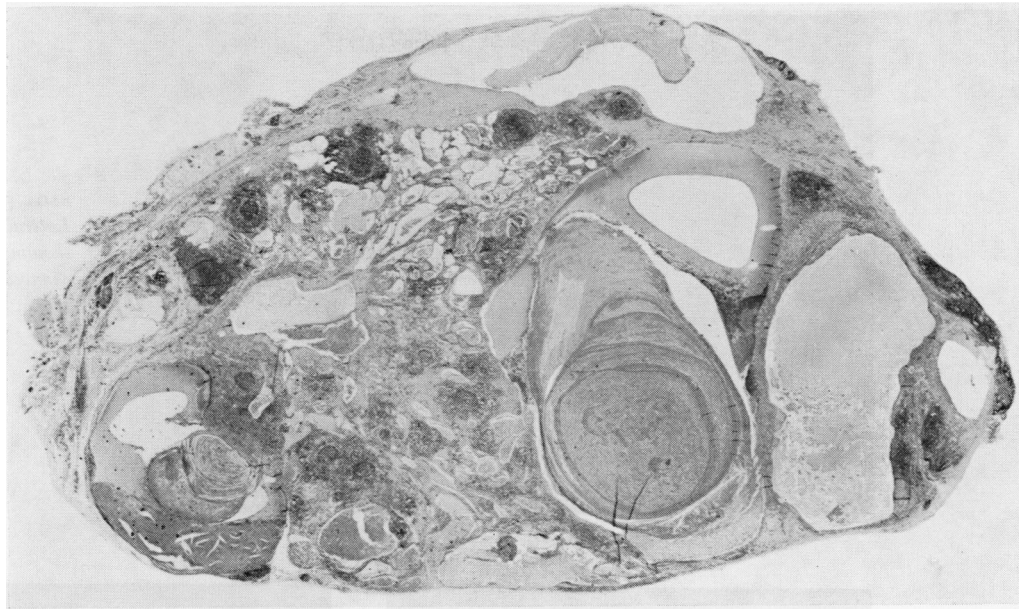

FIG. 3 *Case 3. Lesion composed of dilated venous channels, small arteries, and lymphocytic foci in stroma, some of the latter showing germinal centres. Haematoxylin and eosin.* ×5·5

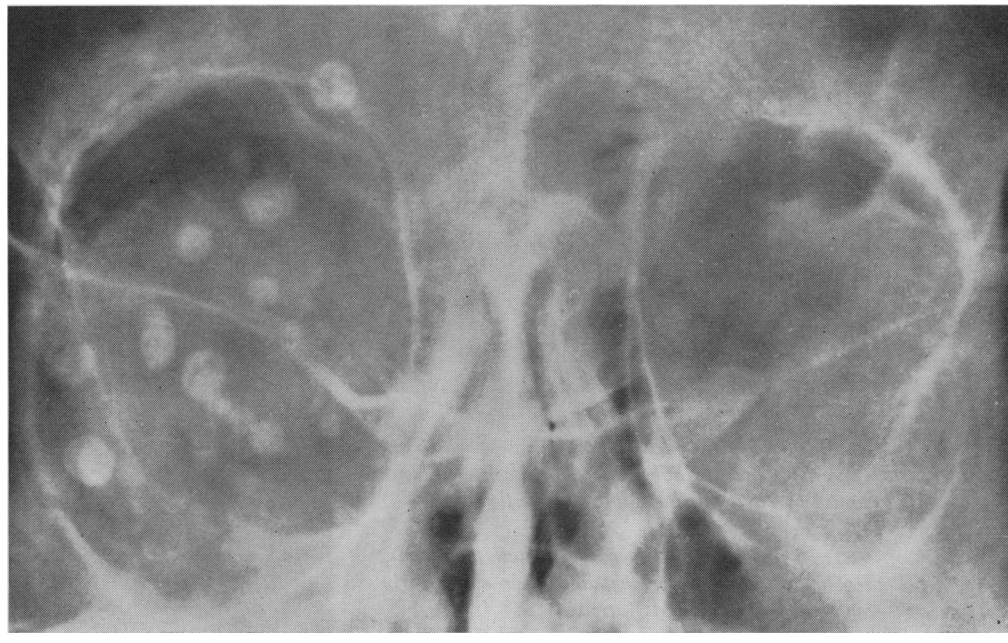

FIG. 4 *Case 4. Radiograph of enlarged right orbit with multiple phleboliths*

1958) and we were able to demonstrate this on orbital venography in a varix in the orbit of a 15-year-old girl (Case 7) (Figs 7 and 8). In this patient contrast remained in the varix outlining its upper and posterior surface some 5 minutes after the time of the original injection into the orbital venous system.

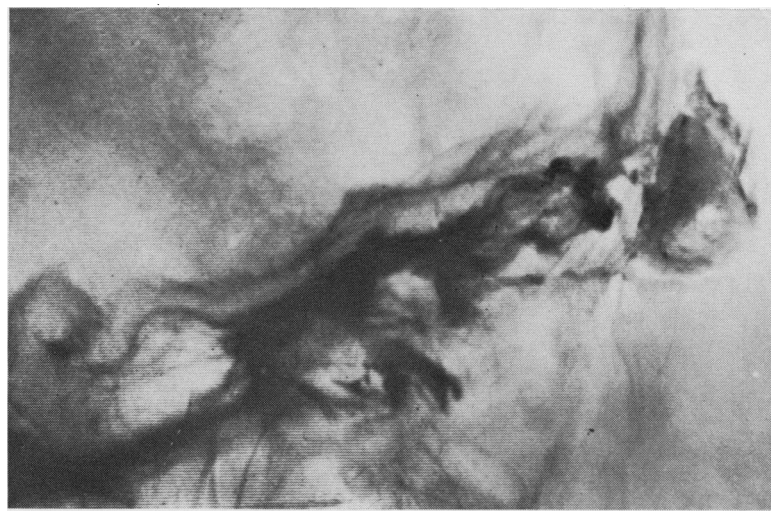

FIG. 5 *Case* 4. *Lateral subtraction film, showing abnormal venous channels in the upper orbit*

(6)

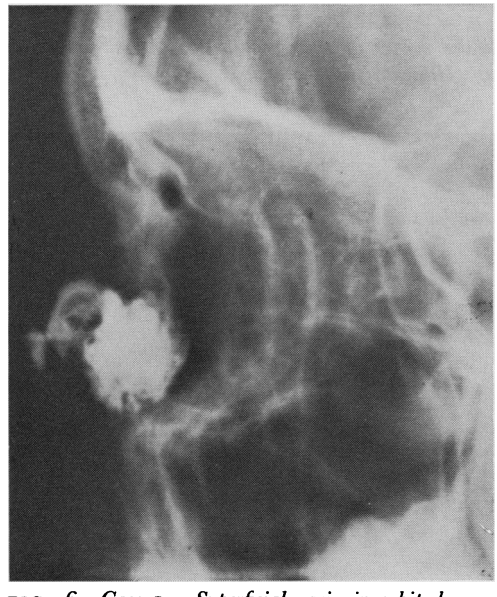

FIG. 6 *Case* 5. *Superficial varix in orbit demonstrated by direct injection (lateral projection)*

(7)

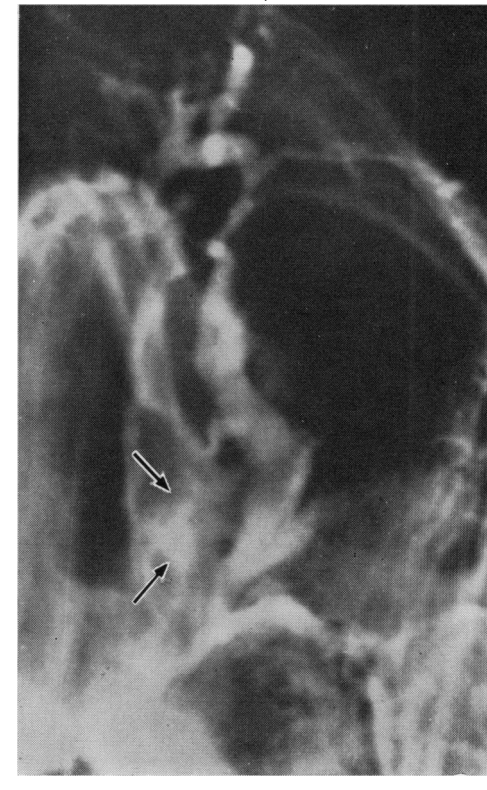

FIG. 7 *Case* 7. *Venogram (axial projection), showing dilated superior ophthalmic vein and a varix situated medially in orbit (arrowed)*

Histopathology

The essential pathological changes were a single dilated venous channel as in Cases 5 and 12 or several dilated venous channels as in Cases 3 and 11 (Figs 3, 11, 12, and 13). Thrombosis of these veins had occurred in Cases 5, 11, and 12 (Fig. 14). In addition, stromal fibrosis had compressed one of the veins in Case 11, and Cases 2 and 3 were charac-

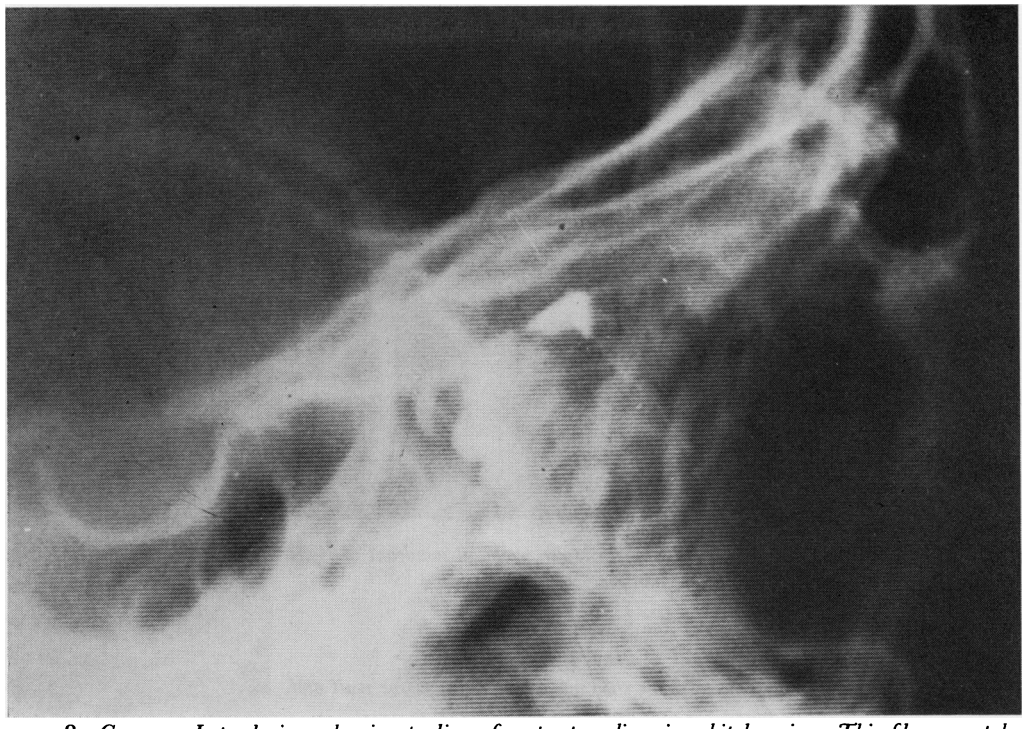

FIG. 8 *Case 7. Lateral view, showing pooling of contrast medium in orbital varix. This film was taken 5 min. after intravenous injection of contrast medium*

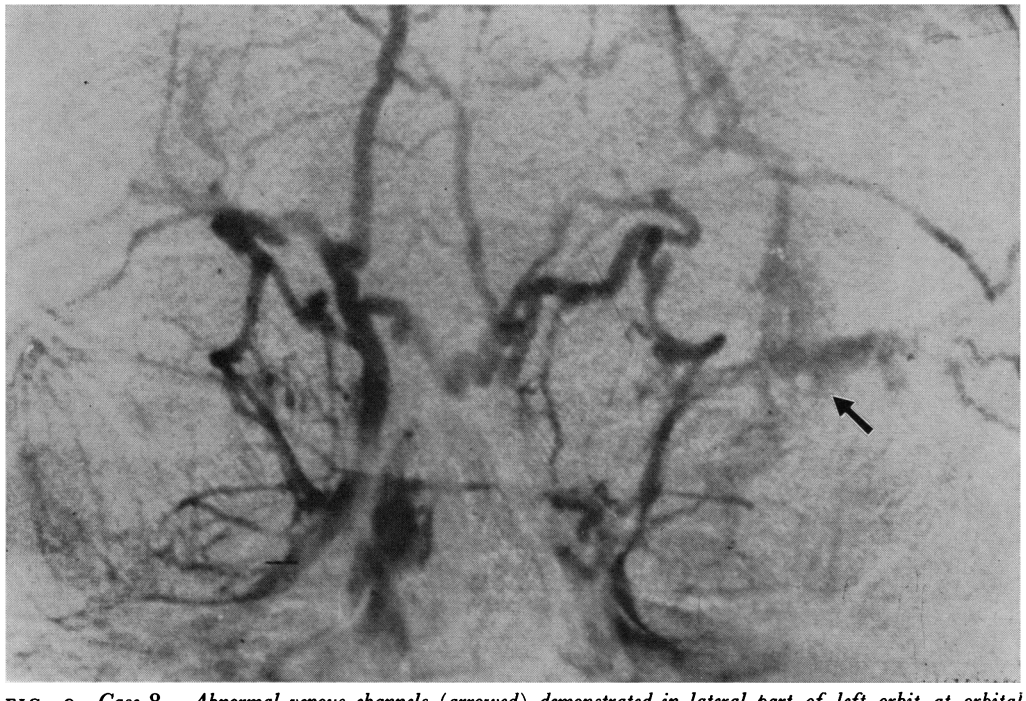

FIG. 9 *Case 8. Abnormal venous channels (arrowed) demonstrated in lateral part of left orbit at orbital venography (subtraction film)*

terized by lymphocytic infiltration of the stroma between the vessels. Although small arteries were seen in Cases 2 and 3, there was no evidence of an arteriovenous communication in any of the five cases.

(10)

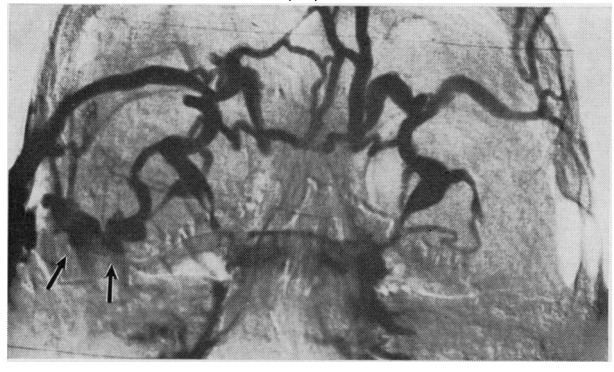

FIG. 10 *Case* 10. *Varices in lower temporal quadrant of right orbit (arrowed) draining into pterygoid plexus*

(11)

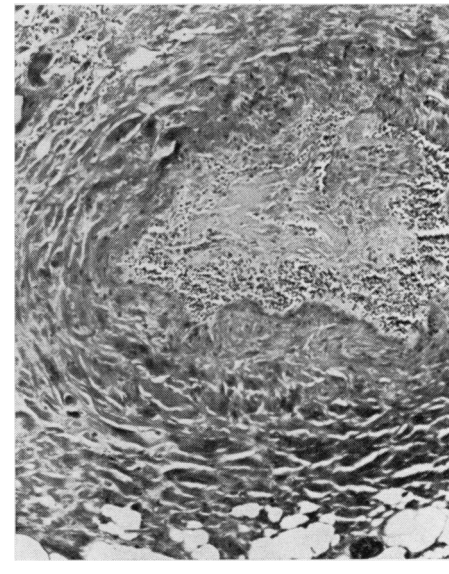

FIG. 11 *Case* 11. *Vein showing a connective tissue wall and sparse elastic fibres. Verhoeff-van Gieson.* ×80

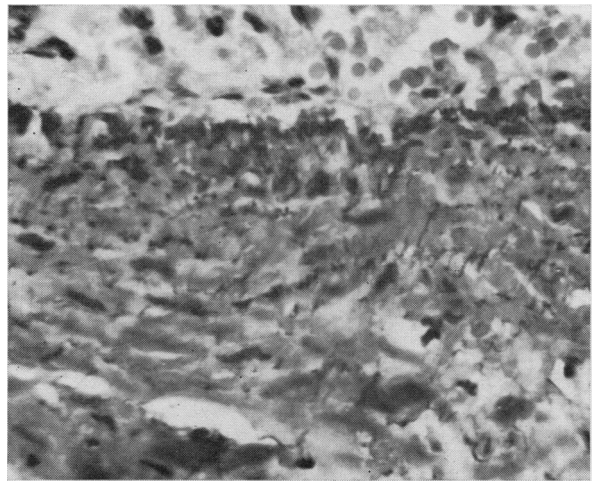

FIG. 12 *Case* 11. *Part of wall of vein, showing internal elastic lamina. Verhoeff-van Gieson.* ×440

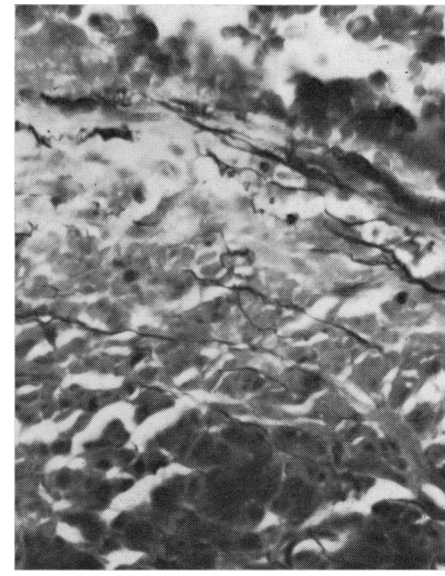

FIG. 13 *Case* 11. *Part of wall of vein, sh sparse elastic fibres. Verhoeff-van Gieson.* ×

Discussion

Early writings concerning orbital varices consist, for the most part, of clinical descriptions of single cases without surgical or pathological proof of the presence of abnormal orbital

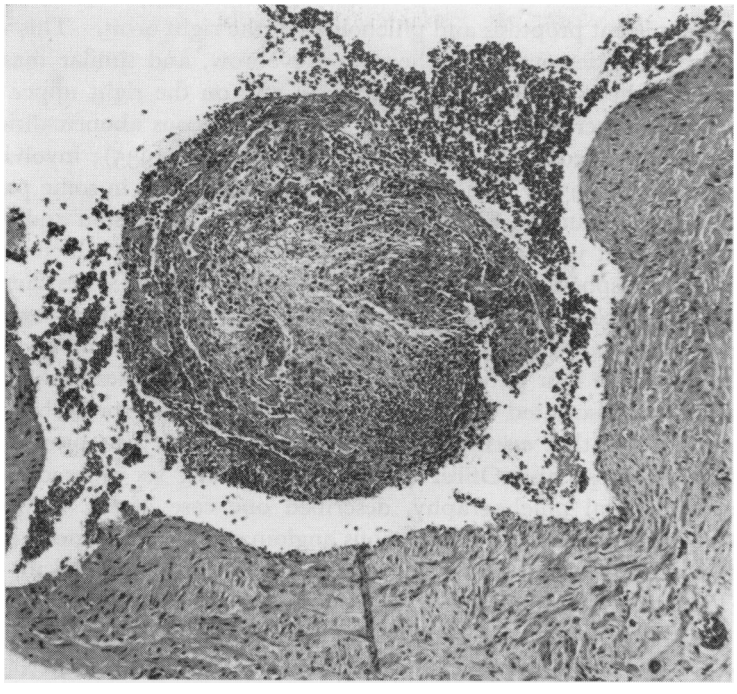

FIG. 14 *Case 12. Portion of wall of vein, containing thrombus. Haematoxylin and eosin.* ×70

veins or of their origin. In these patients the diagnosis of orbital varices was inferred clinically because of the presence of intermittent proptosis, associated in some patients with visibly dilated veins in the anterior orbit. Reports of varices proven at surgery were given by Lowenstein (1911), Germain and Weill (1928), and Rumjanzewa (1930), but in none of these records of operations was there any mention of the possible aetiology of the venous enlargement.

It was not until the report of a case of intermittent proptosis and orbital varices by Walsh and Dandy (1944) that a definite cause was discovered. In this patient the varices were shown, at craniotomy, to be secondary to an arteriovenous malformation in the middle fossa. Duke-Elder (1952), quoting this patient, suggested that this may be the cause in many, if not all such cases. On the other hand, Hobbs, du Boulay, and Davis (1960), reporting a case of cavernous haemangioma, regarded orbital varices as a more fully developed form of haemangioma. It has already been pointed out (Lloyd, 1965) that neither of these theories could account for all types of varix found in the orbit, since these could be either:

(*a*) Primary: due to a congenital venous malformation in the orbit.

(*b*) Secondary to an arteriovenous shunt, either intracranially or within the orbit itself.

CONGENITAL VENOUS MALFORMATION

We believe the twelve patients described belong to this category and form a recognizable clinico-radiological entity distinct from secondary orbital varices. A typical description of the clinical features of this syndrome was given by Magnus (1884) and more recently by Hu Yung-Lin and Shih Chen-Jung (1954). The latter authors recorded a 26-year-old

patient with intermittent proptosis and phleboliths in the right orbit. This was associated with venous abnormalities on the forehead and eyebrow, and similar masses and large varicose veins over the right side of the abdomen and on the right upper arm and leg. Other authors have described patients with associated venous abnormalities outside the orbit: on the buccal mucosa (Gastreich, 1932; Marchesini, 1935); involving the upper and lower limbs (Kira, 1932). The venous malformation may, in some patients, extend into the middle fossa (Hanafee, Shiu, and Dayton, 1968). Brauston and Norton (1963) also described a patient with a venous malformation in the orbit which extended backwards to an enlarged sphenoidal fissure. In four patients in our series there was clinical or radiological evidence of extension outside the orbit, usually involving the scalp and frontal bone on the affected side, but one patient (Case 4) had a venous abnormality of the hard palate. These four patients can be regarded as examples of the full syndrome, *i.e.* orbital varices and associated venous malformations outside the orbit, but in the remaining patients we believe that essentially the same abnormality was present but confined to the orbit. Aron-Rosa and Offret (1967), in recording six cases of orbital varices demonstrated by orbital phlebography, described one case of the Klippel-Trenaunay syndrome, which was associated with a venous angioma of the orbit and eye, and it would seem that the condition we have described in the orbit may be related to, or be part of, the Klippel-Trenaunay syndrome.

SECONDARY ORBITAL VARICES

During the period in which this series was collected, three patients were investigated in the *X* ray Department for orbital varices and were shown to have a carotico-cavernous fistula of non-traumatic origin causing secondary dilatation of the otherwise normal venous system in the orbit. These patients for the most part are easily distinguishable on clinical grounds from cases of primary or congenital varices. Another and clinically less distinguishable cause of secondary dilatation of orbital veins is an arteriovenous malformation, which may be situated either within the orbit itself or intracranially. The first recorded case was that of Walsh and Dandy (1944), who described an 18-year-old girl with intermittent proptosis and dilatation of the intraorbital veins due to an arteriovenous malformation in the middle fossa. More recently de Lima and Penzholz (1968) have published a case in which orbital varices were shown at surgery to be associated with an arteriovenous malformation in the anterior fossa of the skull, lying immediately above the orbital roof, through which the dilated orbital veins communicated directly with the malformation. Examples of arteriovenous malformations within the orbit giving rise to orbital varices are more numerous. Jacas, Ley, and Campillo (1959) published the arteriograms of a 31-year-old patient with intermittent proptosis and orbital varices due to an arteriovenous malformation in the orbit. Intraorbital arteriovenous malformations have also been recorded in the literature by Huber (1951), Krayenbühl (1962), Lombardi (1967), and Aron-Rosa and Offret (1967). In these patients varices may either be dilated normal veins or may consist in part of the venous component of the arteriovenous malformation.

In one patient in this series (Case 3), a large solitary orbital varix was associated with small, multiple intracranial arteriovenous malformations; however, since these were remote from the orbit containing the varix and there was no demonstrable connection between them, it was felt that the vascular anomalies were not causally connected, and that the orbital varix should be classified as a primary venous malformation.

Conclusion

The classification of orbital varices into primary (congenital venous malformation) and secondary (due to an arteriovenous shunt) is in accord with the view expressed by Walsh and Dandy (1944) that:

"It is possible that there are two general types of lesion causing intermittent exophthalmos: (*i*) Arterio-venous with pulsation, and (*ii*) Venous without pulsation."

This statement, though generally true, needs some qualification on the clinical side, since an arteriovenous malformation, even when present within the orbit, may not produce a pulsating exophthalmos, nor indeed is this a constant feature of a carotico-cavernous fistula. On the other hand, pulsating exophthalmos has been reported in a patient with an entirely venous malformation in the orbit by Brauston and Norton (1963), who considered it to be due to an enlargement of the sphenoidal fissure allowing transmission of cerebral pulsation to the orbital contents. It is thus not possible to be certain of the aetiology of orbital varices in a given case on clinical grounds alone, nor are the plain-*x* ray changes pathognomonic, although it would seem that the presence of phleboliths is almost diagnostic of a venous malformation. It is therefore essential to investigate patients by angiography before any treatment is considered.

In the presence of a bruit or pulsating exophthalmos indicating the likelihood of an arteriovenous shunt, carotid arteriography is the method of choice, whereas an intermittent exophthalmos without pulsation or bruit should be investigated in the first instance by orbital venography, either by the frontal vein technique (Vritsios, 1963) or by direct injection of a superficial varix (Fig. 6). The presence of enlarged orbital veins as a cause of proptosis should be recognized if the appropriate contrast studies are undertaken. Frontal venography now enables us to differentiate several types of venous abnormality within the orbit and thus places their treatment on a more rational basis.

Our thanks are due to the surgeons of Moorfields Eye Hospital for allowing us to present their cases, to Prof. Barrie Jones for his help and encouragement, and to Miss R. Rhodes for her secretarial assistance.

References

ARON-ROSA, D., and OFFRET, G. (1967) *Bull. Soc. Ophtal. Fr.*, numéro spécial, "La phlébographie orbitaire: Soc. Ophtal. de Paris. Rapport annuel", p. 85

BRAUSTON, B. B., and NORTON, E. W. D. (1963) *Amer. J. Ophthal.*, **55**, 701

DE LIMA, L. J., and PENZHOLZ, H. (1968) *J. Neurol. Neurosurg. Psychiat.*, **31**, 81

DUKE-ELDER, S. (1952) "Text-book of Ophthalmology", vol. 5, p. 5399. Kimpton, London

GASTREICH, C. (1932) *Klin. Mbl. Augenheilk.*, **88**, 773

GERMAIN, L., and WEILL, G. (1928) *Ann. Oculist. (Paris)*, **165**, 542

HAGER, G. (1958) *v. Graefes Arch. Ophthal.*, **159**, 662

HANAFEE, W. N., SHIU, P. C., and DAYTON, G. O. (1968) *Amer. J. Roentgenol.*, **104**, 29

HOBBS, H. E., DU BOULAY, G., and DAVIS, R. E. (1960) *Brit. J. Ophthal.*, **44**, 551

HUBER, A. (1951) *Bull. Soc. franç. Ophtal.*, **64**, 347

HU YUNG-LIN and SHIH CHEN-JUNG (1954) *Chin. med. J.*, **72**, 482

JACAS, R., LEY, A., and CAMPILLO, D. (1959) *J. Neurol. Neurosurg. Psychiat.*, **22**, 330

KRAYENBÜHL, H. (1962) *J. Neurosurg.*, **19**, 289

KIRA, B. (1932) *Acta Soc. ophthal. jap.*, **36**, 1627 (German Summary, p. 122)

LLOYD, G. A. S. (1965) *Clin. Radiol.*, **16**, 339

LOMBARDI, G. (1967) "Radiology in Neuro-Ophthalmology". Williams and Williams, Baltimore

LÖWENSTEIN, A. (1911) *Klin. Mbl. Augenheilk.*, **49 (2),** 183
MAGNUS, H. (1884) *Ibid.*, **22,** 62
MARCHESINI, E. (1935) *Ann. Ottal.*, **63,** 263
RUMJANZEWA, A. F. (1930) *Z. Augenheilk.*, **71,** 247
WALSH, F. B., and DANDY, W. E. (1944) *Trans. Amer. ophthal. Soc.*, **42,** 334
VRITSIOS, A. (1963) *Arch. Soc. Ophtal. Grèce Nord*, **12,** 223