Nodular fasciitis causing unilateral proptosis

R. H. PERRY,* P. S. RAMANI,† V. McALLISTER,§ R. M. KALBAG,†
C. R. KANAGASUNDARAM‡

*From the Department of Neurosurgery,† the Department of Ophthalmology,‡ and the Departments of
Neuroradiology§ and Neuropathology,* Newcastle General Hospital*

Although nodular fasciitis has been described in relation to the superficial structures of the eye, it has not been described as an intraorbital lesion causing proptosis. We describe a patient with unilateral proptosis caused by intraorbital nodular fasciitis.

Case report

A 34-year-old housewife presented with a 1 year

Address for reprints: P. S. Ramani, Department of Neurosurgery, Newcastle General Hospital, Westgate Road, Newcastle upon Tyne NE4 6BE

history of prominence and puffiness of the left eye. She had no pain or visual disturbance apart from double vision on upward gaze.

EXAMINATION

The abnormal findings were restricted to the left eye. There was a 4 mm proptosis and the eye was also depressed by 3 mm compared with the right eye. There was diplopia on elevation which could not be related to a specific muscle. The visual acuity was normal and the fundi did not show any abnormality. A full blood count, erythrocyte sedimentation rate, and chest x ray were normal.

FIG. 1 Left carotid angiogram. Lateral subtraction (bone free) view showing prominent ophthalmic artery with unusual group of pathological vessels (arrow) in orbit

FIG. 2 *Orbital venogram. Antero-posterior projection showing medial and downward displacement of left superior ophthalmic vein (arrow)*

FIG. 3 *Orbital venogram. Basal view showing medial displacement of left superior ophthalmic vein (arrow)*

SPECIFIC INVESTIGATIONS

X rays of the skull included views of the orbit, optic foramina, and sinuses. No bony or soft tissue abnormality was demonstrated. A left carotid angiogram revealed a prominent ophthalmic artery with an unusual inferior loop in the proximal portion. There were, in addition, pathological irregularities of the vessels in the upper part of the orbit (Fig. 1). An orbital venogram demonstrated a space-occupying lesion in the superior lateral part of the orbit producing downward and medial displacement of the superior ophthalmic vein (Figs 2 and 3). The space-occupying lesion was additionally demonstrated as a circular area of uptake with a technetium 99M gamma camera scan.

TREATMENT

A left frontal craniotomy was made and the orbit was approached by removing its roof. There was an apparently encapsulated vascular lesion, in the superior/lateral portion of the orbit (Fig. 4). It was situated behind the orbital margin outside the muscle cone and had no macroscopic connexion with the orbital wall, the globe, muscles, or nerves. There was no obvious connexion with the lacrimal gland. The main mass of the lesion was situated behind the equator of the globe. The lesion was readily dissected and removed intact without any immediate postoperative complications. The patient made an uneventful recovery.

HISTOPATHOLOGY

Macroscopically the specimen was ovoid and well circumscribed measuring $2\text{ cm} \times 2\text{ cm} \times 1.6\text{ cm}$. It had a firm resilient consistency and a uniform dull grey colour.

Histologically the specimen was predominantly made of fibrocytes. The majority had a spindle or ovoid shape with

a vesicular nucleus and an ill-defined, homogeneous, and slightly eosinophilic cytoplasm. In the central region the cells were arranged as a dense series of interlacing bundles in which were present occasional collagen fibres and intercellular clefts. Towards the periphery the clefts were more readily identifiable as vascular channels and in this region they constituted a predominant feature. Reticulin fibres were abundant (Fig. 5) and were uniformly distributed as a fine network without any characteristic pattern. They demarcated small intercellular spaces often containing red blood cells (Fig. 6). No haemosiderin pigment was demonstrated but a single foreign body type giant cell was identified. Small round cells resembling lymphocytes were scantily distributed in the lesion. In spite of the well circumscribed macroscopic appearance no capsule was present at the periphery of the lesion. Lymphoid aggregates were not present and there were no features to suggest that the lesion had originated in muscle, nerve, the meninges, or the globe.

Although the lesion was strikingly vascular, endothelial lined blood vessels were not common and clefts composed of endothelial cells were not seen. The characteristic feature of a haemangiopericytoma, therefore, was not present. In addition the reticulin pattern was not characteristic of this tumour.

Discussion

The histological features of this lesion can be identified as belonging to the curious fibrovascular proliferative entity termed 'nodular fasciitis'. Font and Zimmermann (1966) described 10 such cases in relation to the superficial structures of the eye (predominantly the eyelids), but none was within the orbital cavity. Tolls, Mohr, and Spencer (1966) at the same time reported a lesion at the insertion of the inferior rectus muscle adherent to the sclera. It was

FIG. 4 Photograph taken from above during operation showing lesion in situ. It was exposed, through left frontal craniotomy, by removing roof of orbit. Lesion looks well encapsulated. Inset: Actual size of lesion after it was removed in one piece

FIG. 5 Photomicrograph of edge of lesion showing prominent reticulin fibres and marked vascularity. Reticulin $\times 40$

FIG. 6 Photomicrograph demonstrating cellular area of fibroblastic proliferation with vascular channels. Note haphazard cellular arrangement with reticulin fibres outlining cells and demarcating vascular spaces. Reticulin $\times 120$

easily seen preoperatively. A further case occurring as a 6 mm nodule under the bulbar conjunctiva was described by Anton (1967), this lesion was apparently

recognized clinically as nodular fasciitis and histological examination merely confirmed the diagnosis. Levitt, de Veer, and Oguzhan (1969), in the last report of this lesion in relation to the orbit, described a 1 cm nodule occurring at the nasal margin of the right orbit.

Examples of nodular fasciitis are well documented in relation to the orbit but no reports to date have indicated that the lesion can occur in the intraorbital tissue planes and cause proptosis. Mortada (1971) described two cases of fibromata adherent to the periosteum of the orbit, but both these tumours had characteristic features distinguishing them from the present lesion. In a description of tumours and tumour-like lesions causing exophthalmos, Tsukahara (1964) described no case of nodular fasciitis. Similarly in the 245 cases of unilateral exophthalmos reported by Bullock and Reeves (1959) and the 61 primary tumours of the orbit described by Lombardi (1967) examples of nodular fasciitis are not described.

In this region the 'pseudotumour of the orbit' links a group of unresolved pathological entities. The histological appearance of the majority of these (lymphocyte aggregates and inflammatory response) would exclude examples of nodular fasciitis. However a small number of 'pseudotumours' with a predominantly fibrovascular appearance may well have been unrecognized examples of nodular fasciitis (Reese, 1956).

Although well recognized as a pathological entity (Konwaler, Keasbey, and Kaplan, 1955; Price,

Silliphant, and Shuman, 1961; Stout, 1960, 1961; Soule, 1962; Dahl, Angervall, Magnusson, and Sterner, 1972) the stimulus which leads to the vascular connective tissue proliferation has not been identified. Trauma has been cited as an aetiological agent but in the present case there was no history of a traumatic incident to the eye apart from an operation for a squint when the patient was nine years old. Multinucleated giant cells are well described in nodular fasciitis. Soule (1962) noted them in 25 out of 56 lesions, remarking that they occurred as 'unobtrusive cells in more cellular zones'. Other authors (Stout, 1961; Font and Zimmerman, 1966; Hutter, Stewart, and Foote, 1962) comment that they occur as non constant features but only Soule states that they are not of foreign body type. In the present case a search of many sections revealed only one multinucleated giant cell and it is of interest that it contained a rectangular eosinophilic hyaline body 0.5 μ m across. It is likely that it represents phagocytosis of degenerate (possibly crystalline) material.

Usually nodular fasciitis appears as an isolated and

rapidly growing mass in the subcutaneous tissue of the upper extremity or trunk. Their cellular appearance and rapid growth have led to their being confused with sarcomatous lesions (Stout, 1961); they are however benign and do not recur after complete excision (Hutter and others, 1962).

This case demonstrated an uncommon site and unusual presentation for nodular fasciitis. Carotid angiography, by demonstrating a vascular tumour, should enable further examples to be identified preoperatively.

Summary

A case report of an unusual case of nodular fasciitis in the orbit presenting with unilateral proptosis is described, and the radiological features are outlined. The histological features are discussed and the benign nature of the lesion stressed. Nodular fasciitis arising in the orbit and presenting as unilateral proptosis has not previously been reported in the literature.

References

- ANTON, M. (1967) *Čs. Oftal.*, **23**, 450
 BULLOCK, L. J., and REEVES, R. J. (1959) *Amer. J. Roentgenol.*, **82**, 290
 DAHL, I., ANGERVALL, L., MAGNUSSON, S., and STERNER, B. (1972) *Path. europ.*, **7**, 211
 FONT, R. L., and ZIMMERMANN, L. E. (1966) *Arch. Ophthalm.*, **75**, 475
 HUTTER, R. V. P., STEWART, F. W., and FOOTE, F. W. (1962) *Cancer (Philad.)*, **15**, 992
 KONWALER, B. E., KEASBEY, L., and KAPLAN, L. (1955) *Amer. J. clin. Path.*, **25**, 241
 LEVITT, J. M., DE VEER, J. A., and OGUZHAN, M. C. (1969) *Arch. Ophthalm.*, **81**, 235
 LOMBARDI, G. (1967) *Neuro-ophthal.*, **6**, 111
 MORTADA, A. (1971) *Brit. J. Ophthalm.*, **55**, 350
 PRICE, E. B., SILLIPHANT, W. M., and SHUMAN, R. (1961) *Amer. J. clin. Path.*, **35**, 122
 REESE, A. B. (1956) 'Tumors of the Eye and Adnexa.' US War Dept, Surgeon General's Office, Atlas of Tumor Pathology, Sect. 10, 38, Washington
 SOULE, E. H. (1962) *Arch. Path.*, **73**, 437
 STOUT, A. P. (1960) *Minnesota Med.*, **43**, 455
 ——— (1961) *Cancer (Philad.)*, **14**, 1216
 TOLLS, R. E., MOHR, S., and SPENCER, W. H. (1966) *Arch. Ophthalm.*, **75**, 482
 TSUKAHARA, I. (1964) *Jap. J. clin. Ophthalm. (Tokyo)*, **18**, 693