Figure 1 Case 1. (A) Fluorescein angiogram of right eye. Visual acuity was 20/200. There was a 2 disc diameter subfoveal pigment epithelial detachment with smooth borders. The juxtapapillary choroidal neovascular membrane was located inferotemporally adjacent to the optic nerve. There was a distance of less than 1 disc diameter between the edge of the choroidal neovascular membrane and the edge of the retinal pigment epithelial detachment. (B) Fluorescein angiogram of the same eye, 5 weeks after treatment. Visual acuity was 20/20 and the retinal pigment epithelial detachment had resolved.

improvement of visual acuity.^{2,3} We successfully treated two patients who had serous subfoveal pigment epithelial detachments and juxtapapillary choroidal neovascular membranes. The juxtapapillary choroidal neovascular membranes were extrafoveal and were not contiguous with the retinal pigment epithelial detachments. In addition, the areas of laser photocoagulation were not contiguous with the retinal pigment epithelial detachments.

CASE 1

A 73-year-old woman presented with decreased vision in her right eye for 1 week. Visual acuity was 20/200 right eye, 20/25 left eye. She had subretinal fluid, haemorrhage, and exudate on the temporal margin of the optic disc and a subfoveal pigment epithelial detachment in her right eye. Fluorescein angiography demonstrated a juxtapapillary choroidal neovascular membrane and a 2 disc diameter subfoveal pigment epithelial detachment in her right eye (Fig 1A).

The juxtapapillary choroidal neovascular membrane was treated with the argon green laser. Three weeks after treatment, the retinal pigment epithelial detachment was flat and the patient's visual acuity was 20/20 (Fig 1B). At her last examination, 2 years after laser treatment, her visual acuity was 20/20. She had no signs of recurrence.

CASE 2

An 82-year-old woman noted decreased visual acuity in her left eye for 2 months. Her visual acuity was 20/40 right eye and 20/200 left eye. She had subretinal fluid, haemorrhage, and exudate on the temporal margin of the optic disc and a subfoveal pigment epithelial detachment in her left eye. A fluorescein angiogram showed a juxtapapillary choroidal neovascular membrane and a subfoveal pig-

Figure 2 Case 2. (A) Fluorescein angiogram of left eye. Visual acuity was 20/200. The retinal pigment epithelial detachment under the fovea is hyperfluorescent as is the juxtapapillary choroidal neovascular membrane. (B) Fluorescein angiogram of the left eye 3 weeks after laser photocoagulation. Visual acuity was 20/20 and the retinal pigment epithelial detachment had resolved.

ment epithelial detachment in her left eye (Fig 2A). The juxtapapillary choroidal neovascular membrane was treated with argon green laser. Three weeks later, the retinal pigment epithelial detachment was flat, and her visual acuity was 20/50 (Fig 2B). Her last examination was 2 years after the laser treatment. Her visual acuity was 20/25. She had no signs of recurrence.

COMMENT

The juxtapapillary choroidal neovascular membranes in these two patients were clinically and angiographically distant from the subfoveal serous retinal pigment epithelial detachments. There are three possible ways that laser photocoagulation of a juxtapapillary choroidal neovascular membrane could cause flattening of a distant subfoveal pigment epithelial detachment. Firstly, and most likely, the choroidal neovascular membrane may have been leaking fluid under the non-detached retinal pigment epithelium with enough force to cause a distant retinal pigment epithelial detachment. Photocoagulation of the choroidal neovascular membrane could ablate the source of the subretinal pigment epithelial fluid and cause rapid resolution of the subfoveal pigment epithelial detachment. Secondly, there may have been an occult connection between the choroidal neovascular membrane and the retinal pigment epithelial detachment that was not detectable angiographically. The feeder vessels to such an occult area of choroidal neovascular membrane may have been obliterated by the laser treatment causing the retinal pigment epithelial detachment to flatten. Finally, the retinal pigment epithelial detachment may have arisen independent of the choroidal neovascular membrane. The area of the laser scar may have allowed egress of sufficient fluid from the subretinal pigment epithe-

lial space into the subretinal or choroidal space for the pigment epithelial detachment to flatten.

We recommend laser photocoagulation in patients with extrafoveal choroidal neovascular membranes and central visual acuity loss from subfoveal retinal pigment epithelial detachments even if the two lesions are neither clinically nor angiographically connected.

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Linear scleroderma with orbital involvement: follow up and magnetic resonance imaging

EDITOR.—Scleroderma is a chronic connective tissue disease of unknown aetiology.¹ Linear scleroderma is an uncommon dermatological disorder which produces an unilateral and localised band-like linear atrophy of the skin and its underlying structures.

Orbital involvement has been described, but is very rare.^{1,2}

CASE REPORT

A 21-year-old woman first presented to an ophthalmologist because of a progressive narrowing of the right palpebral fissure over a period of 3 years with loss of lashes of the eyelids. Further ophthalmic examination was normal. An orbital computed tomogram (CT) was normal.

Three years later there was additional enophthalmia and an entropion of the lower right eyelid was seen (Fig 1). Visual acuity was 8/10 in the right eye and 10/10 in the left. Visual field examination and colour vision were normal.

With the exception of a localised area of parietotemporal alopecia on the right side, no other skin defects could be detected. General examination revealed no muscular pain, no arthralgia, no fatigue, and no other systemic symptoms. Laboratory examination of muscle enzymes, tissue antibodies and complement

Figure 1 Right eye: note the narrowing of the palpebral fissure and the loss of lashes of the eyelids.

factors was normal. There were no inflammatory factors and thyroid tests were within normal limits.

On an orbital CT the right eye was displaced laterally by a band-like mass. The medial rectus muscle and oblique superior muscle could not be identified. Muscle and conjunctival biopsy only showed some oedema in these structures.

The presumed diagnosis of localised scleroderma was confirmed by a dermatological examination. Initially no treatment was established.

An orbital magnetic resonance image (MRI) confirmed the CT findings but also demonstrated that the right superior rectus muscle was involved in the mass (Fig 2A and B). A new biopsy only showed muscular and fibroid tissue without signs of inflammation.

Two years after the first admission the patient complained of increasing right sided headache, photophobia, and progressive decrease of the visual acuity on the right side from 8/10 to 3/10. Visual field examination and colour vision were normal. Biomicroscopic examination showed a punctate keratitis on the right side. Funduscopy revealed a hyperaemic optic disc. Follow up MRI showed a progressive increase in the size of the orbital mass with extension into the apex. The optic nerve could only be identified following intravenous gadopentetate dimeglumine injection (Fig 2C–F). A canthotomy and an eyelid plasty were performed for restoration of the right upper eyelid (Fig 3).

The unilateral loss of vision progressed slowly to finger counting at 1 metre the following year. The visual fields showed normal peripheral limits, but colour vision was absent in the right eye. Cortisone therapy (Medrol, 32 mg/day) and radiotherapy (4 Gy on the right orbit, followed by 20 Gy in fractions of 2 Gy) were initiated, but the patient complained of increasing pain in and around the right eye. The patient became blind on the right side. Ophthalmic examination showed a mydriatic non-reactive pupil and a pale optic disc on the right side. Two follow up MR examinations did not show a further progression of the orbital abnormalities.

COMMENT

Scleroderma may occur as a systemic disease or as a localised form. Localised scleroderma presents in three clinical forms: generalised, morphoea (atrophic and sclerotic skin lesions), and linear scleroderma. In the linear form the lesions are arranged in a band-like linear distribution and may involve and fix the deeper layers of the skin and underlying structures. If it involves the frontoparietal region it is called scleroderma 'en coup de sabre' because of the depression that extends below the level of the skin. In patients with morphoea or linear scleroderma, the disease progresses slowly and may last from a few months to many years, but the prognosis is worst in the en coup de sabre type. According to some authors, it may lead to homolateral facial hemiatrophy (Parry–Romberg syndrome).¹ However, the relation between linear scleroderma en coup de sabre and Parry–Romberg syndrome remains unclear.³

Olver *et al* have reported a patient with bilateral acquired Brown's syndrome and combined lichen sclerosis et atrophicus and morphoea.⁴ They suggest that the deep subdermal fibrosis may limit the passive movement of the superior oblique tendon. The clinical features of Brown's syndrome

Figure 2 (A,B). Coronal T1 weighted MRI (TR/TE = 520/15 ms). The superior rectus muscle, the superior oblique muscle, and the medial rectus muscle on the right side appear enlarged and cannot be distinguished from each other (A) (arrowheads). The orbital apex is not involved (B) (arrow). (C) Axial T2 weighted MRI (TR/TE = 2200/80 ms). (D, E) Coronal T1 weighted MRI (TR/TE = 500/15 ms). (F) Coronal T1 weighted MRI after gadopentetate dimeglumine injection (TR/TE = 500/15 ms). The orbital abnormalities return a low signal on T2 weighted images (C) (arrows). The apex is involved and the entire orbital fat is infiltrated (D,E). Note that the optic nerve is only visible after the contrast administration (arrow) (F).

were not present in our patient, but can be seen in localised scleroderma.

The cutaneous pathological features depend upon the stage of the disease: the striking accumulation of inflammatory cells among collagen bundles of the lower dermis and among the fat cells and collagen bundles of the subcutaneous tissue is followed by the replacement of the subcutaneous tissue by hyalinised connective tissue.¹

Involvement of the orbit and the eye is uncommon but has been described.^{1,2} Involvement of the extrinsic eye muscles is extremely rare and only CT findings have been reported.²

The clinical findings in this case are typical of localised linear scleroderma: the age and sex of the patient, the slow progression, the distribution and nature of the superficial defects, and the unsuccessful therapy. This patient has a significant induration of the scalp but there is no extension below the forehead and is therefore not considered as a Parry–Romberg syndrome. The term pseudotumour is misleading and inappropriate because of the presence of enophthalmia and the absence of mass effect.^{5,6} The latter can be explained by the atrophy of orbital fat that is only partially replaced by fibrosis. Orbital fat atrophy is a basic mechanism of enophthalmia, and its relation to linear scleroderma has already been reported.^{7,8} The thickening of the orbital muscles in this case can probably be secondary to orbital fat atrophy and fibrosis.²

Although the imaging findings are not specific, important information can be ob-

tained. A precise localisation of the orbital abnormalities, a clinicoradiological correlation and the follow up of the abnormalities can be achieved.

The CT findings are non-specific.⁹ In this patient the lesion is of the chronic and infiltrative or diffuse type. It does not invade or distort the globe and there is no bone erosion.^{5,6}

MRI findings in patients with a non-specific orbital inflammatory condition have been described as hypointense to fat and isointense to muscle on T1 weighted images. On T2 weighted images the abnormalities were isointense or slightly hyperintense to fat in nine out of 10 cases.¹⁰ We found similar signal characteristics in this patient. These appearances were markedly different from those in other diseases, including malignancy and haematoma, which appeared markedly hyperintense to fat on T2 weighted images.¹⁰ The use of gadopentetate dimeglumine was helpful in assessing the compression by the lesion on the optic nerve.

To our knowledge, this is the first report on follow up of localised linear scleroderma by MRI. The abnormalities on MRI correlate with the clinical progression. MRI might be better to distinguish between orbital fat atrophy, oedema, and fibrosis but more experience is needed to know if MRI can distinguish the type and the activity of an inflammatory condition.

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Figure 3 Clinical photograph taken after the two minor surgical procedures shows the progression of the disease.

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Primary orbital fibrosarcoma developing in the scleral stroma

EDITOR,—Fibrosarcomas are thought to be malignant tumours developing from fibroblasts.¹ Primary fibrosarcoma of the orbit is very rare. For example, only five cases were reported among a total of 986 orbital tumours in adults.² Recently we encountered the case of primary orbital fibrosarcoma attached to the posterior pole of the globe, the posterior wall of which was encapsulated with fibrous tissue similar to the sclera, although the tumour had infiltrated the posterior pole of the globe and the optic nerve sheath. The fibrosarcoma in this case is thought to have developed in the scleral stroma.

CASE REPORT

A 56-year-old woman was first referred to our clinic for examination of an intraconal tumour of the left eye. On admission her visual acuity was 20/20 in the right eye and 2/200 in the left eye, with correction. There was no record of previous radiation therapy and no history of orbital injury. The main ophthalmoscopic findings were the presence of marked choroidal folds, disc oedema, and opticociliary shunt vessels in the left eye. Systematic and haematological investigations revealed no abnormalities. Results of computed tomography of the orbit showed a relatively well defined intraorbital mass attached to the posterior pole of the globe (Fig 1). In axial T1 weighted magnetic resonance images, the posterior wall of the tumour was thought to be encapsulated (Fig 2). The tumour was not invasive towards the retrobulbar space; however, it had partially infiltrated the scleral stroma. The posterior pole of the globe was compressed and flattened by the tumour. In sagittal T1 weighted images, it was observed to compress the optic nerve; its boundary towards the optic nerve sheath was partially blurred. Pathologically, the tumour was composed of interlacing bands of spindle-shaped cells forming a herringbone pattern (Fig 3). The number of mitoses was three per ten high-power fields ($\times 450$). Immunohistochemical staining for vimentin was positive, indicating that the tumour had originated from mesodermal cells. The tumour was infiltrated into the sclera and into the optic nerve sheath. Up to about 1 year after the exenteration, no recurrence of the tumour had been detected.

Figure 1 Axial plain (right) and enhanced (left) computed tomographic scans of the orbit. Arrowheads indicate calcification in the tumour.

Figure 2 Magnetic resonance images of the orbit. Axial T1 image (top left) shows that the posterior wall of the tumour was encapsulated and that the tumour compressed and flattened the posterior pole of the globe. The boundary of the tumour towards the globe was irregular and partially blurred. The tumour was enhanced homogeneously with gadolinium-DTPA (top right; axial T1 image with fat suppression). The tumour had compressed the optic nerve sheath as well as the globe (bottom left; sagittal T1 image). The intensity of the tumour is inhomogeneous in T2 image (bottom right; coronal T2 image).

Figure 3 Photomicrographs of the tumour. Spindle-shaped cells arranged in characteristic herringbone pattern (left; haematoxylin and eosin, $\times 120$). Immunohistochemical staining of the tumour for vimentin was positive (middle; $\times 120$). The right panel shows that tumour cells (upper part of the section) had infiltrated the sclera (lower part of the section) (haematoxylin and eosin, $\times 40$).