The Surgical Management of Vasoproliferative Tumours

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Key Words

Vasoproliferative tumours · Transconjunctival cryotherapy · Perforating diathermy · Transscleral resection · Plaque radiotherapy · Transvitreal resection

Abstract

Vasoproliferative tumours consist of glial and vascular proliferations, most commonly found in the inferotemporal quadrant. The need for surgery arises when there is diagnostic uncertainty or sight-threatening complications are present. A variety of surgical techniques have been used in the management of vasoproliferative tumours and are discussed in this review.

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Background

Vasoproliferative tumours consist of glial and vascular proliferations most commonly found in the inferotemporal quadrant [1]. They are thought to arise from within the sensory retina [2] and can be idiopathic (74%) or arise secondarily to a number of disease processes [1, 3]. The clinical manifestations are well described [1]. Most vasoproliferative tumours are solitary, with a yellow-pinkish

colour and may be surrounded by intraretinal haemorrhage, intraretinal or subretinal exudate and hyperpigmentation of the retinal pigment epithelium (fig. 1) [4]. The average age at diagnosis for solitary tumours is 40 years, but diffuse vasoproliferative tumours present at a much younger age [1]. Diagnosis is based on the clinical appearance of the tumour, with ancillary tests such as fluorescein angiography being of less use due to the anterior location of most tumours [3]. Difficulty in establishing a diagnosis may arise when the tumour has an atypical appearance or location and can simulate retinal capillary haemangioblastomas, eccentric choroidal neovascularisation and choroidal melanomas [3–5].

A recent histopathological report [2] has suggested a link between the pathobiology of proliferative vitreoretinopathy (PVR) and vasoproliferative tumours. Retinal pigment epithelial cells were noted to be present in both the tumours and the associated epiretinal and subretinal membranes. As the authors noted, periretinal membranes containing retinal pigment epithelial cells are the pathological hallmark of PVR, and they hypothesised a link between PVR and vasoproliferative tumours. Certainly it is a common clinical observation that epiretinal membranes associated with vasoproliferative tumours tend to be rapidly contractile.

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Fig. 1. Vasoproliferative tumour.

Vasoproliferative tumours are associated with a number of sight-threatening complications, most commonly cystoid macular oedema, vitreous haemorrhage, macular epiretinal membranes and progressive subretinal exudation threatening the macular region [1, 6]. The optimum management of vasoproliferative tumours is unknown due to their rarity and uncertainty regarding their underlying pathobiology. In the majority of cases observation alone is required [1]. Medical management includes laser photocoagulation or photodynamic therapy for smaller tumours [7], and bevacizumab for associated macular oedema [8]. The need for surgery arises when there is diagnostic uncertainty or sight-threatening complications are present. However, some authors advocate earlier intervention when the tumours are small, allowing treatment before any sight-threatening complications develop [4, 9]. A variety of surgical techniques have been used in the management of vasoproliferative tumours and they are described below.

Perforating Diathermy

Perforating diathermy coagulation is a historical technique initially suggested in 1930 [10] and used as an effective treatment for large peripheral retinal angiomas [11] until the introduction of photocoagulation and cryotherapy. The technique involved placing one ring of diathermy around the edges of the tumour, which had been delineated by indirect ophthalmoscopy. A 3-mm diathermy was then passed through the sclera into the centre of the tumour. The depth of coagulating perforations was monitored by indirect ophthalmoscopy to ensure there was no accidental perforation of the retina. Multiple perforations were made until the tumour showed uniform whiteness [11].

Transconjunctival Cryotherapy

Transconjunctival cryotherapy has been widely used in the management of vasoproliferative tumours. Fifty-five percent of patients within the original case series on the classification and clinical manifestations of vasoproliferative tumours underwent cryotherapy as a treatment of choice [1]. A standard triple freeze-thaw cycle is used. Cryotherapy is recommended for tumours with an apical diameter of <2 mm as larger tumours are difficult to treat in one session due to their thickness [4, 9]. Repeat treatments with cryotherapy can be successful in the management of larger tumours but may also increase the risk of further exudative changes [4] and can lead to complete exudative retinal detachment [12].

Plaque Radiotherapy

Both ^{125}I and ^{106}Ru radiotherapy plaques have been used in the management of vasoproliferative tumours. In a case series containing 30 vasoproliferative tumours (23 with associated exudative detachment) treated with ^{125}I plaque radiotherapy, Cohen et al. [13] reported successful treatment, with 29 of 30 tumours regressing and 15 of 23 exudative detachments resolving completely. Visual acuity remained stable or improved in 22 (73%) of the 30 cases. The plaques were placed using the standard surgical technique [14], with tumours being selected for treatment if their thickness was greater than 2.5 mm. Similar results were reported in a case series of 35 symptomatic tumours treated with ^{106}Ru plaque therapy [15]. The mean tumour thickness was 2.8 mm and exudative reti-

nal detachment was present in 25 of the cases. Visual acuity remained stable or improved in 57% of the cases.

Both series report a relatively low rate of radiation-induced complications. However, the better visual outcome within the ^{125}I group may be accounted for by the lower mean dose of radiation when compared with the ^{106}Ru group (mean radiation dose of 108 Gy at the tumour apex and of 416 Gy at the base in the ruthenium group compared with 40 Gy and 110 Gy in the iodine group, respectively) [13].

Transscleral Resection

Local transscleral resection of vasoproliferative tumours has been described in a small number of patients [16]. These patients presented with moderately large tumours associated with extensive exudative detachments, making it difficult to exclude the possibility of choroidal melanoma. The technique involves creating a lamellar scleral flap over the tumour. The tumour is then dissected away from the retina and removed along with the deep scleral lamella. Sulphur hexafluoride is injected as an endotamponade. During the procedure a systolic blood pressure of 50 mm Hg must be maintained [16, 17]. In cases of diagnostic difficulty this procedure may save the eye from enucleation and provides tissue for histological analysis [16, 18], albeit with the local and systemic risks of the procedure.

Pars Plana Vitrectomy Combined with Endocryotherapy and/or Endolaser

Shields et al. [1] listed endocryotherapy as a possible treatment modality within the original case series on the classification and clinical manifestations of vasoproliferative tumours. This method of treatment has not been widely used and there is no further literature on the subject. The authors used a confluent long-duration 532-nm endolaser on the surface of 2 medium-sized tumours (with base diameters of approx. 3 mm in each case and thicknesses of 2 and 3.5 mm, but with limited serous detachment) during vitrectomy and membrane peeling procedures for associated macular epiretinal membranes. The laser was applied as adjunctive treatment combined with externally applied cryotherapy and resulted in successful tumour control and no short-term increase in exudation as may have occurred with cryotherapy alone.

Transvitreal Endoresection

Transvitreal endoresection of vasoproliferative tumours has been reported in a small case series [19]. Three patients with exudative macular changes and tumours resistant to conventional treatment with cryotherapy or plaque radiotherapy were selected for the technique. A standard 20-gauge pars plana vitrectomy was performed, with detachment of the posterior hyaloid face. After trimming of the vitreous base and membrane peels, diathermy was applied to all tumour vessels. Endoresection of the tumour was done using the vitrector with endodiathermy to maintain haemostasis. Silicone oil endotamponade was used [19].

Two of the 3 patients within this series underwent simultaneous cataract extraction and lens implantation, both developed severe fibrinous anterior uveitis and pupillary membranes that enveloped the lens implant. Both subsequently had the lens removed and an artisan lens placed. All the patients had an improvement in visual acuity, with no recurrence of the tumour, and resolution of the macular changes.

A similar recently reported technique allows endoresection of the tumour for histological analysis [5]. It can be used when there is diagnostic difficulty, such as in the reported case, where exclusion of a von Hippel-Lindau haemangioblastoma was required. In the case described, vitrectomy was performed with subsequent 360-degree laser around the periphery of the tumour. The tumour was then removed via an enlarged supertemporal sclerostomy after endodiathermy to any feeder vessels and surrounding retinectomy. An air-fluid gas exchange was then performed, followed by perfluoropropane instillation [5]. The patient regained vision of 6/12 with no recurrence of tumours.

Discussion

Vasoproliferative tumours are uncommon lesions of the peripheral retina that are thought to be reactive rather than neoplastic in nature [2]. The majority of tumours require only observation unless sight-threatening complications are present or there is diagnostic uncertainty.

The surgical techniques are varied, with no agreement as to the best method for the management of these tumours. Transconjunctival cryotherapy is the most widely used technique for the treatment of tumours with a thickness of <2 mm. This is an effective treatment, but care should be taken as any associated exudation may change

for the worse if repeated treatments are used. Plaque radiotherapy is an effective treatment for larger tumours, especially those with significant exudative detachment, or tumours that are not responsive to transconjunctival cryotherapy. Iodine and ruthenium plaques have both shown promising results with few radiation-induced complications.

Diagnostic uncertainty most commonly arises when there are multiple tumours, the tumour is in an atypical position or extensive exudative changes make identification difficult. Referral to a specialised oncology service is recommended as transscleral resection may allow tumour removal and spare the patient an unnecessary enucleation.

The more recently described vitreoretinal approaches show promising results. They enable the surgeon to deal with any associated complications such as vitreous haemorrhage or epiretinal membranes whilst removing the tumour. Although technically demanding, these techniques may offer good results in patients who have tumours resistant to standard treatments.

Disclosure Statement

Both J.S. and D.S. declare they have no conflicts of interest.

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