Retinoblastoma – trends in conservative management

Retinoblastoma kills in the main by extension along the optic nerve, gaining access to the subarachnoid space and the meninges. Prognosis for survival therefore is related to the age at diagnosis, and in those countries with ready access to medical services this age is now, on average, 12 months for bilateral cases and 24 months for unilateral cases. Unilateral cases therefore have a slightly worse prognosis in the early years (second non-ocular tumours make the longer term survival for the bilateral cases much worse), but for both groups 5 year survival rates of around 90% are regularly reported.

Retinoblastoma is usually managed by enucleation but there is growing agreement that treatment must be individualised to the specific patient, and that the only absolute indication for enucleation is the possibility of optic nerve involvement. An increasing trend towards conservative management is being seen, with Shields and his group1 reporting a steady reduction in the proportion of patients having an eye removed for retinoblastoma from 96% in 1974–8 to 75% in the period 1984–8. This reduction applied to both unilateral and bilateral cases.

Abramson et al2 report a similar although less marked trend with a statistically significant reduction in the use of primary enucleation from 97% to 87% for unilateral cases and from 97% to 91% for bilateral cases, comparing the periods 1951–65 and 1966–80 (a drop from 67% to 58% of eyes removed in the bilateral cases). Neither study reports any increase in the approximate 5% chance of metastatic retinoblastoma in patients treated conservatively as opposed to those treated with enucleation, although both studies are at great pains to point out the continuing importance of enucleation as a common, safe, and effective way to manage unilateral and bilateral retinoblastoma.

The mainstay of conservative therapy is external beam radiotherapy which has been used since 1903 to treat this radiosensitive tumour. Other conservative methods of treatment include photocoagulation, cryotherapy, and radioactive plaques where the isotopes have included cobalt-60, ruthenium-106, iridium-192, and iodine-125. These are often used in conjunction with external beam therapy. Shields’s group3 reports on 400 patients of whom 103 had a plaque at some stage of their treatment, although it was the initial therapy in only 31.

There are two reports in this issue of the journal (pp 109 and 112) giving the experience of the team at St Bartholomew’s Hospital using external beam radiotherapy either by whole eye irradiation or using a lens sparing technique. This approach reflects the desire to tailor the treatment to the individual patient and to minimise where possible the side effects of radiation. Treating the whole eye gives a tumoricidal dose to the whole retina, but invariably produced cataract in the Barts patients within 2 years. While increased fractionation of the dose of radiation from an initial 9–10 fractions to an eventual 20 fractions reduces posterior segment complications such as radiation chorioretinopathy and optic neuropathy, the anterior segment complications remain severe. Damage to lids, cornea, and lacrimal gland tissue reduces the options in aphakia correction and may itself produce significant corneal damage with reduced vision. The technique was successful on its own in 57% of their cases, while the addition of focal therapy boosted the success rate to 80%, the remaining 20% of eyes having to be enucleated (three of these because of the development of neovascular glaucoma secondary to radiation induced retinal ischaemia and not because of any active tumour).

Lens sparing external beam radiotherapy, using Harnett’s modification of Shipper’s technique4 has been used at Barts since 1986 to treat selected cases of retinoblastoma that fall mainly into Reese-Ellsworth groups I to III. This is because the lens sparing technique delivers an inadequate dose of radiation to the peripheral retina so that one is confined to treating posteriorly placed tumour, or alternatively one can destroy tumour in the anterior retina using focal means and then treat the remainder with the lateral beam. Twenty one per cent of the treated eyes in the Barts series required prior focal treatment to anterior tumour. Looking only at Reese-Ellsworth groups I–III, a 92% success rate was achieved by the lens sparing method compared with an 85% success rate using whole eye radiation. Only one cataract developed, and that is attributed to the use of an anteriorly placed plaque, while no significant anterior segment morbidity was seen in the lens sparing group.

New tumours developed in 19% of the lens sparing group, however, compared with 5% of the whole eye group, and most of these were anterior to the equator (92%), whereas only 22% of new tumours in the whole eye group were anterior to the equator. The risk of these anteriorly placed tumours constitutes the main obvious problem with the newer technique, and demands close monitoring, while the technique itself requires considerable expertise in its administration. These factors simply strengthen the argument for a few specialist teams dealing with all retinoblastomas at a national level.

We await the development of methods which will reduce our dependence on radiation which is reported to increase the risk of mortality from a second neoplasm in those individuals with the genetically determined form of the disease, where the cumulative probability of death from a second primary neoplasm is reported to reach 26% at 40 years.6

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