Ocular Tumours

Lesions are not always what they seem

by Howard Larkin in Milan

A day after being kicked in the face by his sister, a seven-year-old boy presented with redness and swelling below his left eye. With negative ocular and medical histories, haematoma seemed the most likely diagnosis. But Jerry A Shields MD, director of the ocular oncology service at Willis Eye Institute, Philadelphia, US, thought the 3.5cm x 2.0cm lump suspicious, so he excised it.

Microscopic examination of the circumscribed mass he removed revealed malignant elongated cells with cross striations. It was rhabdomyosarcoma, the most common malignant orbital tumour seen in children, Dr Shields said in his keynote address to the 2nd World Congress of Paediatric Ophthalmology and Strabismus.

Rhabdomyosarcoma can metastasise to the lungs, lymph nodes and other organs, Dr Shields noted. As recently as the 1970s it had an 80 per cent to 90 per cent mortality rate, but with radiation and chemotherapy, the cure rate today is over 90 per cent – if it is caught in time.

“Rhabdomyosarcomas can occur on the eyelid, they can masquerade as trauma, and you must keep that in mind,” he cautioned.

But appearances can just as easily deceive in the other direction, Dr Shields added. In another case, an eight-month-old boy presented with a lump on his left upper lid that had been enlarging for five weeks. Physical exam and MRI findings suggested it might be rhabdomyosarcoma, so the mass was removed.

This time, the child required no cancer treatment. Pathology revealed fascicles of spindle cells and mitotic figures, leading to a diagnosis of nodular fasciitis.

“It can resemble rhabdomyosarcoma but it is entirely benign and will spontaneously regress. But you need a good pathologist to distinguish them,” said Dr Shields, who spoke on what’s new and interesting in the area of paediatric ocular tumours and pseudotumours.

Adult cancer in children

Sometimes tumours seen in adults show up in children, Dr Shields noted. He presented a case in which a mass in the left upper lid of a 10-year-old girl turned out to be adenosid cystic carcinoma, a highly malignant tumour of the lacrimal glands.

“It generally occurs in adults, but actually has a biphasic age distribution and is seen in young children in their first or second decade.”

Historically, adenoid cystic carcinoma has a dismal prognosis and the textbooks recommend orbital extirpation, Dr Shields said. But he questioned if such drastic surgery should be done on a child with 20/20 vision and possibly no residual tumour.

As an option, Dr Shields has had success with orbital plaque brachytherapy. However, the radioactive seeds are placed on the outside of the protective shield so they mainly irradiate the orbit, where microscopic residual is more likely, rather than on the inside, as is typical for treating lesions in the globe.

Retinal astrocytic hamartomas related to tuberous sclerosis complex are thought to be stable with few complications, requiring minimal treatment and follow-up, Dr Shields said. However, he has treated several cases where the lesions were quite aggressive.

A 10-year-old boy presented with a blind, painful right eye. A mass was visible that produced ultrasound echoes suggesting retinoblastoma, Dr Shields said. The eye showed calcification and a total retinal detachment, and was enucleated. But the dissected lesion did not look like retinoblastoma, Dr Shields said. It contained large glial cells, gemiostotic astrocytes and calcospheres, which reflected ultrasound energy.

The final diagnosis? Giant cell astrocytic tumour of the retina associated with tuberous sclerosis.

“It is identical to the brain tumours in children with tuberous sclerosis. We have seen several more. In three cases, each had a mass that looked like retinoblastoma causing retinal detachment,” Dr Shields said.

These findings suggest a re-evaluation of astrocytic hamartomas of TSC may be in order, Dr Shields said.

“This is one of these supposed benign stable lesions that filled the retina, grew out and filled the vitreous cavity and broke out through the limbus producing an extracocular extension. So I wonder now if the astrocytic hamartomas of tuberous sclerosis are all that stable. We are seeing now that some are very aggressive and they may even require enucleation.”

Affecting about 10 per cent of newborns, capillary haemangiomas of the eyelids and orbit are one of the most commonly seen paediatric tumours, Dr Shields noted.

Complications of extensive tumours can include amblyopia and strabismus. Traditional management includes resection, observation, patching and injected or oral corticosteroids. However, while corticosteroids generally are effective, complications can include retinal vascular obstruction, necrosis of the eyelid, subcutaneous fat atrophy and skin de-pigmentation for injections, and adrenal suppression for oral delivery, Dr Shields noted.


“They do regress by themselves, but this certainly speeds it up.”