The speciality of Oncology has benefitted from development of niche sub specialities. These specialists bring with them the knowledge of their parent specialities and add a knowledge of comprehensive Cancer Care to the betterment of the patient.

The field of Ophthalmic plastic surgery and ocular oncology is one such area.

As this Newsletter will showcase, patients of ocular and ocular adnexal malignancies need the care of specialists who understand the disease process and treatment, with a specific knowledge of the organ involved.

It is hoped that this will provoke a healthy academic discourse, and I congratulate the department for this pioneering academic effort in this new speciality.

Dr. B.S. Ajai Kumar,
Chairman, HCG
A Case Report

Management of Eyelid Sarcomatoid Squamous Cell Carcinoma (SCC): A Rare Spindle Cell Variant.

Case discussion: A 65 year old male patient presented with a mass on the right upper eyelid. The lesion was clinically diagnosed as Pyogenic granuloma and surgery attempted in another hospital. Due to uncontrolled bleeding, the surgery was stopped midway and a specimen of the mass was sent for Histopathological examination (HPE). It was diagnosed as Rhabdomyosarcoma and referred to our centre for further management. Magnetic resonance Imaging and Positron Emission Tomography revealed that the mass was confined to the right upper eyelid and limited by the orbital septum.

Excision biopsy of right upper eyelid mass with frozen section control was planned. The mass was excised and after confirmation of negative margins, eyelid reconstruction was performed.

The HPE revealed Spindle cells consistent with sarcoma like tumour on light microscopy. Immunohistochemistry was performed in which the tumour cells expressed CK, CK5/6, p63 and were negative for Vimentin, EMA, Myf-4, S-100 and Melan A. A diagnosis of Spindle cell variant of SCC was made. Post operative Brachytherapy was given to counter micrometastasis.

Conclusion: Sarcomatoid or spindle cell variant of SCC is relatively rare variant of SCC. Immunohistochemistry helps to differentiate when there is a confusing HPE picture, helping make the correct diagnosis as well as guiding treatment.
Retinoblastoma: A Success Story

Retinoblastoma is a cancer of the eye. The cancer starts in the retina — the light sensing part of the eyeball on the inside of your eye.

Retinoblastoma is usually a cancer of young children, but in rare cases even adults can suffer from this cancer. It is the most common eye cancer of childhood. Incidence may vary from 1/18,000 births in developed nations to 1/12,000 births in developing nations.

It can be heritable as well as non-heritable. The non-heritable type is the most common type accounting to about 90-93% of all retinoblastomas.

What makes Retinoblastoma curable?

Reasons for high success rate are many

Genetic basis of the tumour being identified

1. Early diagnosis of the tumour
2. Multiple treatment options
3. Cutting edge basic and clinical research

Genetic Basis

At the beginning of the 20th century the survival rate of children with Retinoblastoma was less than 17%. After modest success for many decades the discovery of the Rb1 gene mutation (1986) was game changing. More than 70 years ago, it was recognized that retinoblastoma sometimes has a genetic basis and that the pattern (when genetic) is classic Mendelian autosomal dominant. It was not until Alfred Knudson proposed his two-hit hypothesis and the subsequent molecular confirmation of his calculation that it was recognized that this autosomal-dominant pattern is caused by the loss of a normally occurring gene that is now referred to as the RB1. Although it was originally thought to be of importance only in retinal cancers, it is now recognized that loss of the normal RB1 gene is an important step in cancer development in most adult non ocular cancers.

Early Diagnosis

Early detection of Retinoblastoma is the key.

Leukocoria - which translates to white pupil - in other words whitish appearance from the centre of the eye is the most important sign of Retinoblastoma, which brings it to the attention of the parents to seek help.

There are other signs too but Leucocoria is the most common sign. This early sign and hence early diagnosis is also responsible for high cure rates seen in Retinoblastoma.

Treatment Options: The biggest advantage in the treatment of Retinoblastoma is that it is responsive to multiple options of treatment unlike other tumours.

The characteristics of the tumour is

1. Extremely chemosensitive and hence it has high response to chemotherapy
2. Radiosensitive, so Radiotherapy is an option even when the tumour has spread outside the eyeball.
3. It is amenable various local therapy like Laser therapy, thermal therapy, cryotherapy and plaque brachytherapy. This is possible because we can visualize the tumour using an ophthalmoscope.
4. When only one eye is involved, Surgery - Enucleation can sometimes give permanent cure from the cancer.

Pioneering research

As it stands today, in the best centers in the world for Retinoblastoma care, the success rates are staggering:

- 95% Life salvage
- 90% Eye salvage
- 85% Vision salvage
**Issue in focus**

**Retinoblastoma: A Success Story**

This is staggering because just 50 years ago, the rates of survival in children with retinoblastoma was less than 30% even in the best centers. Today Retinoblastoma is treated using a team based approach. The team consists of an Ocular Oncologist, Pediatric Oncologist, Radiation Oncologist, Pathologist, Interventional radiologist and an Ocularist. This approach has resulted in better care for children with Retinoblastoma.

References:


5. Retinoblastoma: Recent Update and Management Frontiers.


Ocular surface squamous neoplasia (OSSN) comprises a wide spectrum of dysplastic alterations of the squamous epithelium of the surface of the eye, e.g., the cornea and the conjunctiva ranging from "precancerous" lesions to bona fide invasive carcinoma. In the former case, they are classified as carcinoma-in-situ lesions in conjunctival-corneal intra-epithelial neoplasia (CCIN) and in the later in invasive squamous cell carcinoma (SCC). The clinical presentation of Ocular surface squamous neoplasia (OSSN) varies across a wide spectrum and is classified based on the degree of epithelial and stromal infiltration. The epithelial infiltration can range from mild to severe dysplasia to full-thickness epithelial dysplasia (carcinoma in situ) and invasive squamous cell carcinoma, when tumor cells invade through the epithelial basement membrane and into the conjunctival and/or corneal stroma. Ocular surface squamous neoplasia (OSSN) can involve the conjunctiva or the cornea individually but more commonly start in the conjunctiva and extend across the limbus to involve the adjacent cornea.

Surgical excision is the traditional therapy for OSSN. Surgical excision involves excision of the lesion with wide surgical margins. Surgery can be followed by adjunctive cryotherapy to reduce the recurrence rate.

Recurrence rates after surgical excision have been reported as high as 33% with clear surgical margins and of up to 56% with positive surgical margins. Due to the reported high recurrence rates, adjunctive medical interventions for OSSN have been proposed. Local medical therapy has the advantage of treating the entire ocular surface, avoiding wide excision, which poses the risk of stem cell deficiency and long-term ocular surface problems. Mitomycin C, 5-fluorouracil, and interferon alpha 2b have been found to be effective in the management of OSSN. IFNa2b drops are well tolerated and have minimal side effects.
Multidisciplinary Case Discussion

The eighteen year old orbital tumour.

An 18yr old man presented to us with a mass in the right eye which has been slowly growing since birth. This mass was biopsied twice in 1998 and 2003 and on both occasions HPE was vascular harmartoma. A repeat biopsy was done which was again confirmed the tumour to be a vascular harmartoma.

We discussed this in our tumour board and it was decided that we excise the tumour in its entirety. So we planned an Exenteration and excision biopsy of all surrounding lesions.

Since we had a tissue diagnosis of a benign tumour we performed a lid sparing exenteration. We also excised the other extraorbital spillover tumours. We transposed a flap from the superficial temporal fascia to cover the inner lining of the orbit and closed the wound by primary suturing. This gave the patient an acceptable cosmesis. He shall be fitted with a skin colour silicone facial prosthesis to achieve further visual symmetry and cosmesis.
Journal Scan


Swathi Kaliki, M.D.*, Anuradha Ayyar, M.D.*, Akshay G Nair, M.D.*, Dilip K Mishra, M.D.†, Vijay Anand P Reddy,*, and Milind N Naik, M.D.*

*Institute for Eye Cancer; and †Ophthalmic Pathology Service, L V Prasad Eye Institute, Hyderabad, India

The above study is a retrospective case series of sebaceous gland carcinoma of eyelid where neoadjuvant chemotherapy was given. The purpose of the above study was to report the efficacy of neoadjuvant systemic chemotherapy in the management of eyelid sebaceous gland carcinoma (SGC).

The mean percentage reduction of tumor basal diameter after neoadjuvant chemotherapy was 74% (median, 80%; range, 30% to 100%). None of them had any major systemic side-effects of neoadjuvant chemotherapy. Postchemotherapy, surgical treatment for residual tumor was performed in 7 cases. Five cases underwent excision biopsy and 2 cases with residual orbital component underwent eyelid-sparing orbital exenteration. No tumor recurrence was noted in any of the 7 cases at a mean follow-up period of 18 months (median, 14 months; range, 3 to 63 months). One patient died due to systemic metastasis.

**Editorial comments:** Neoadjuvant systemic chemotherapy has been found to be effective and safe in the management of eyelid SGC. However in view of the small sample of cases in the above case series it would merit more studies to establish a protocol for neoadjuvant chemotherapy in SGC. This study adds to are already expanding knowledge about the most common type of eyelid cancer in the Indian sub-continent (upto 60% of all eyelid carcinomas are SGC).

Major review

1. Orbital lymphaticovenous malformations: Current and future treatments Nariman Nassiri, MD, MPH, Jack Rootman, MD, FRCSC, Daniel B. Rootman, MD, MSc, Robert A. Goldberg, MD* Division of Orbital and Ophthalmic Plastic Surgery, Stein Eye Institute, David Geffen School of Medicine, University of California at Los Angeles, Los Angeles, California, USA

Orbital lymphaticovenous malformations consist of abnormal vascular channels lined by endothelial cells with a spectrum from venous to lymphatic characteristics. They may bevenous-dominant or lymphatic-dominant. These lesions continue to present management challenges. Total excision or obliteration is not always achievable, recrudescence is common, and interventions carry a risk of damaging normal structures. Patients likely benefit most from a multidisciplinary approach, including both surgical and nonsurgical (e.g., sclerosants and liquid polymers) therapeutic modalities. Targeted biologic therapy would be ideal; nevertheless, this goal is complicated by the heterogeneous venouslylymphatic and stromal characteristics of these lesions. Ideally, antiangiogenic agents targeting both lymphatic and blood vascular endothelial cells will be developed to treat these lesions and reduce their regrowth. Further studies are warranted to enhance our understanding of these orbital lesions with regard to their angiogenic (proliferative) activities and profiles of marker expression, with a goal to produce effective medical therapies.

**Editorial Comments:** Treatment of Orbital lymphaticovenous malformations can be challenging and there is considerable controversy in classification of such lesions and hence the treatment of such. These lesions can cause functional and structural impairments. Patients benefit most from a multidisciplinary approach including both limited invasive procedures and surgical modalities. Because these lesions are typically composed of a spectrum of lymphatic and blood vascular endothelial cells, antiangiogenic agents targeting both lymphatic and blood vascular endothelial cells may hold some promise: however, further studies of angiogenic activity and profiles of marker expression, in carefully classified groups of these lesions, are necessary.

ii. Epibulbar schwannoma in a 12-year-old boy: A case report and review of literature Akshay Gopinathan Nair, Swathi Kaliki, Dilip Kumar Mishra1, Tarjani Vivek Dave, Milind N Naik, Institute for Eye Cancer, 1Department of Ophthalmic Pathology, L V Prasad Eye Institute, Hyderabad, Telangana, India

A healthy 12-year-old boy presented with complaints of a slow growing painless conjunctival mass of 3 years’ duration in the left eye. Conjunctival/epibulbar schwannoma is an uncommon tumor. In a retrospective, non-interventional case series of 1045 consecutive patients with conjunctival tumors by Shields et al., there were no cases of conjunctival/epibulbar schwannoma. In a large review of 2455 conjunctival tumors by Grossniklaus et al., a single case of conjunctival schwannoma was reported in a 61-year-old male.

**Editorial comments:** Conjunctival or Epibulbar schwannoma is certainly a very uncommon tumour it emphasizes the importance of sending even the benign lesions for histopathological examination. This paper provides good review of literature for this uncommon tumour.
Department of Ophthalmic Plastic Surgery and Ocular Oncology.

Editor: Dr. Raghuraj Hegde
MS Ophthalmology (RIO- Kolkata), FAICO (Oculoplastic Surgery). Fellowship Orbit, Ophthalmic Plastic Surgery and Ocular Oncology (NUHS, Singapore)
Consultant, Orbit, Ophthalmic Plastic Surgery & Ocular Oncology - HCG Oncology Hospitals, Bangalore.
Email id: raghuraj.hegde@gmail.com

Editorial Advisor:
Dr. Ravi Nayar – Dean Academics, HCG

Editorial Assistant:
Mr. Naveen.S – Executive Academics, HCG

Artist
Rama Suresh

Title
Untitled

Media
Mixed Media on canvas

Size
72 x 36 inches

Year
2009

Courtesy
Swasti Art Gallery, HCG K.R.Road
Proceeds of the sale of this painting will be used for assisting poor patients.

For more information contact:
infor@hcgfoundation.org