

**SCIENTIFIC SESSION 4
BRIEF CASE REPORT**

Shilpakalavedika Convention Center
Saturday, January 24, 2004
4:00 PM – 6:00 PM

*Chair: Ralph Eagle
Moderator: Santosh Honavar*

	Presenter	Title of Presentation	Time
1	Hatem Crema	Conjunctival Large Hemangioma in a Case of Presumed Organoid Nevus Syndrome	4:00 PM
2	Ekta Moda	A Giant Epibulbar Dermoid in a Newborn	4:04 PM
3	Uma Chaluvadi	Minimal Manipulation Surgical Technique for Conjunctival Papillomata	4:08 PM
4	Seppo Tuomaala	Evidence-Based Indication for Sentinel Lymph Node Biopsy in a 24-year-old Woman with Conjunctival Melanoma	4:12 PM
5	Anjali Upponi-Patil	A Predominantly Corneal Epibulbar Malignant Melanoma	4:16 PM
6	Sabyasachi Pattanayak	Rhabdomyosarcoma of the Conjunctiva - A Rare Presentation	4:20 PM
7	Milind Naik	Primary Adenoid Cystic Carcinoma of the Eyelid Mimicking a Sebaceous Gland Carcinoma : A Case Report	4:24 PM
8	Ramesh Murthy	Neoadjuvant Chemotherapy in the Management of Sebaceous Gland Carcinoma	4:28 PM
9	Dipansu Basu-Chaudhury	Fibrous Histiocytoma of the Eyelid	4:32 PM
10	Ritu Arora	Malignant Fibrous Histiocytoma of the Orbit - Management Approach	4:36 PM
11	Dipankar Das	An Unusual Case of Retrobulbar Arachnoid Cyst and its Management	4:40 PM
12	Tero Kivela	A Cystic Tumor of the Orbit in an Unborn Baby	4:44 PM
13	Rob De Keizer	Embryonal Rhabdomyosarcoma - An Antenatal Detected Orbital Tumor	4:48 PM
14	Patrick De Potter	Primary Orbital Melanoma Treated with Iodine 125 Plaque Radiotherapy	4:52 PM
15	Raman Mittal	A Case Report of Unknown Orbital Tumor	4:56 PM
16	Ebrahim Shirzadeh	Giant Chondroid Syringoma of the Upper Nasal Orbital Rim : First-Reported Case	5:00 PM
17	Thomas Cummings	Glioblastoma of the Optic Nerve and Chiasm	5:04 PM
18	Vikas Mittal	Pleomorphic Adenoma of Lacrimal Gland in 7-year-old boy : A Case Report	5:08 PM
19	Subhash Betharia	Basal Cell Adenocarcinoma of the Lacrimal Gland - A Newly Recognized Rare Tumor	5:12 PM
20	Harish Pathak	Primary Ductal Adenocarcinoma of the Lacrimal Gland : A Rare Entity	5:16 PM
21	Nagendra Shah	Solitary Plasmocytoma of Orbit	5:20 PM
22	Gaurav Bahl	Bilateral Orbital Lymphoma - A Rare Presentation of Multicentric Synchronous Extranodal MALT Lymphoma	5:24 PM
23	Ashwin Mallipatna	Bilateral Orbital Non-Hodgkin's Lymphoma in a Patient with Acquired Immunodeficiency Syndrome - A Case Report and Review of Literature	5:28 PM
24	Lai Yuen	Sclerosing Extramedullary Hematopoietic Tumor in the Orbit	5:32 PM

CONJUNCTIVAL LARGE HEMANGIOMA IN A CASE OF PRESUMED ORGANOID NEVUS SYNDROME

Hatem Krema

Magrabi Eye Hospital, Cairo, Egypt

To demonstrate an unusual case of a child, with manifestations suggestive of organoid nevus syndrome, who developed a rapidly growing, large conjunctival hemangioma. A mentally retarded male child, born with multiple neurological, cutaneous and ocular lesions, affecting one side of the head, developed a large protruding conjunctival tumor in the ipsilateral eye. Total excision of the mass was done. Histopathology showed cavernous hemangioma-like pattern with fibrous tissue core. Conjunctival hemangioma may be an ocular finding in cases of organoid nevus syndrome.

A GIANT EPIBULBAR DERMOID IN A NEWBORN

Ekta Moda, Nibaran Gangopadhyay

LV Prasad Eye Institute, Hyderabad, India

We report a successful cosmetic and visual outcome of a large epibulbar dermoid in a new born A 40-day-old baby was presented with an unsightly (15mm x 15mm) pedunculated bulbous mass obscuring completely right cornea with full thickness involvement confirmed by immersion B-scan. Excellent cosmesis was achieved by excision of the dermoid along with a full thickness corneal graft. After the failure of first graft, visual rehabilitation was reinforced by a repeat keratoplasty and aggressive amblyopia therapy thereafter. Clear graft with optimal vision is maintained after 15 months. Satisfactory cosmesis with good functional vision were achieved in this rare entity through difficult and serial surgical interventions.

MINIMAL MANIPULATION SURGICAL TECHNIQUE FOR CONJUNCTIVAL PAPILLOMATA

Uma Chaluvadi

Indiana University School of Medicine, Indianapolis, MN, USA

Conjunctival papillomata tend to recur after conventional therapeutic modalities such as simple surgical excision, cryotherapy and even expensive modalities such as Interferon therapy. A relatively inexpensive, safe and highly effective surgical technique combining minimal manipulation surgical excision with Cryo followed by Mitomycin-C application is presented with photographic documentation of cases.

EVIDENCE-BASED INDICATION FOR SENTINEL LYMPH NODE BIOPSY IN A 24-YEAR OLD WOMAN WITH CONJUNCTIVAL MELANOMA

Seppo Tuomaala, Tero Kivela

Helsinki University Central Hospital, Department of Ophthalmology, Helsinki, Finland

A 24-year-old woman presented with three darkly pigmented tumors on the conjunctiva of her right eye that were connected by areas of primary acquired melanosis. She had first noticed dark pigmentation on the area of the caruncle 6 months earlier. Since then the pigmentation had spread rapidly, and a tumor had grown from the caruncle all the way to the upper lacrimal punctum. Clinically, the diameter of the caruncular tumor was 10 mm, and another tumor with a diameter of 8 mm originated from the upper fornix. A third, smaller tumor was seen temporally on the upper tarsal conjunctiva. Because of a statistically high risk of metastasis, a sentinel lymph node

biopsy was performed. Localization of the sentinel nodes was technically successful, and the nodes were negative. Even in high-risk patients with conjunctival melanoma, one predicts two thirds of sentinel lymph node biopsies to be negative.

A "PREDOMINANTLY CORNEAL" EPIBULBAR MALIGNANT MELANOMA

Anjali Upponi-Patil, Geeta Vemuganti, Milind Naik, Santosh Honavar

LV Prasad Eye Institute, Hyderabad, India

We report a rare case of epibulbar malignant melanoma involving the cornea. A 45-year-old female presented with a brown epibulbar mass of 2 months duration. Immersion B-scan ultrasonography and ultrasound bio-microscopy revealed its epibulbar nature. Corneal stroma, anterior segment structures and regional lymph nodes were not involved. The patient underwent wide excision biopsy, alcohol keratoepitheliectomy and conjunctival edge cryotherapy with amniotic membrane transplantation under general anesthesia. Histopathologic examination revealed malignant melanoma of the cornea with involvement of the surrounding perilimbal conjunctiva. Systemic work-up did not reveal any metastases. Corneal melanoma is rare, and can be excised easily. Its predominantly corneal location raises concern about its cell of origin.

RHABDOMYOSARCOMA OF CONJUNCTIVA - A RARE PRESENTATION

Sabyasachi Pattanayak, G. Nagewar, Kaumudee Pattanaik
JPM Rotary Eye Hospital and Research Institute, Cuttack, India

Rhabdomyosarcoma is the most common orbital malignancy in children. Rhabdomyosarcoma presenting as an isolated tarsal conjunctival mass is extremely rare. An 11-year-old girl presented with a well-circumscribed mass of 20-mm diameter of one-year duration in the nasal aspect of upper tarsal conjunctiva. The mass was globular, with irregular surface and firm consistency. She was reported to have a similar lesion in the same location a year ago. It had been resected elsewhere but histopathology report was not available. There was no proptosis, no restriction of ocular motility or regional lymphadenopathy. The tumor was resected. Histopathology revealed embryonal rhabdomyosarcoma. The child underwent external beam radiotherapy and is currently tumor-free.

PRIMARY ADENOID CYSTIC CARCINOMA OF THE EYELID MIMICKING A SEBACEOUS GLAND CARCINOMA: A CASE REPORT

Milind Naik, Santosh Honavar, Geeta Vemuganti

LV Prasad Eye Institute, Hyderabad, India

A 38-year-old female presented with a recurrent nodular tarsus based mass over the right upper eyelid. Regional lymph nodes were not involved. Excision biopsy with lid reconstruction was performed with a clinical diagnosis of sebaceous gland carcinoma. Histopathologic examination revealed an adenoid cystic carcinoma with perineural invasion. Primary cutaneous adenoid cystic carcinoma, an uncommon tumor of the skin, should be added to the differential diagnosis of eyelid tumors.

NEOADJUVANT CHEMOTHERAPY IN THE MANAGEMENT OF SEBACEOUS GLAND CARCINOMA

Ramesh Murthy, Santosh Honavar, Vijay Anand Reddy, Milind Naik, Suryasnata Rath, Raman Mittal

Ocular Oncology Service, LV Prasad Eye Institute, Hyderabad, India

We describe a patient with sebaceous gland carcinoma (SGC) with regional lymph node metastasis treated with neoadjuvant chemotherapy. A 55-year-old lady with recurrent SGC of the left lower eyelid with orbital extension and regional lymph node metastasis was treated with initial neoadjuvant chemotherapy. Eyelid sparing orbital exenteration was done after 3 cycles followed by radiotherapy to the regional lymph nodes. Significant eyelid and orbital tumor volume reduction was achieved with neoadjuvant chemotherapy making partial eyelid-sparing orbital exenteration possible. Lymph node metastasis clinically regressed and helped avoid radical neck dissection. The patient was free of local and regional disease at 1-year follow-up. Neoadjuvant chemotherapy used for chemoreduction could well be a useful modality in the management of SGC.

FIBROUS HISTIOCYTOMA OF THE EYELID

Dipansu Basu Chaudhuri, Pranab, Debasis Bairagi, Sayan Das, Swarup Pathak, Samir Banerjee

Susrut Eye Foundation and Research Center, Kolkata, India

A 68-year-old male presented with a history of recurrent upper lid mass for 23 years for which he was operated twice, last surgery being done 8 years back. Ophthalmological examination showed a multilobulated, firm to hard, relatively well defined mass with surface pigmentation, ulceration and hemorrhage on the right upper eyelid overhanging lower eyelid. Left eye showed mild posterior sub-capsular cataract. CT scan orbit showed right lid mass with involvement of anterior orbit. Total excision of the tumor with skin graft taken from right thigh was done under general anesthesia. The patient was followed regularly up to 3 months. Postoperatively right eye BCVA was 6/12 & left eye BCVA was 6/9. There was partial ptosis in right eye with lid lag and lagophthalmos initially but ptosis was improved subsequently, cosmetically the lid appeared almost normal. The excised tumor was a nodular mass measuring 8-cm X 5 cm X 2 cm with lobulated appearance and grayish yellow homogenous cut surface. Histopathologically tumor was a benign fibrous histiocytoma from skin of the eyelid. To the best of our knowledge only one primary eyelid fibrous histiocytoma has been reported in literature previously.

MALIGNANT FIBROUS HISTIOCYTOMA OF THE CONJUNCTIVA- MANAGEMENT APPROACH

Ritu Arora, Dinesh Kumar Mehta, Usha Raina, Sumit Monga, Lakshmi Bansal, Anuj Gogi

Guru Nanak Eye Center, Maulana Azad Medical College, New Delhi, India

PURPOSE: To report a case of malignant fibrous histiocytoma of the conjunctiva and report its management. **METHODS:** Interventional case report. A 51-year old man presented with an atypical conjunctival mass. It was diagnosed as malignant fibrous histiocytoma on histopathological and immunohistochemical evaluation comprising of the excised specimen. There occurred recurrence of malignancy three months later. The conjunctival lesion was excised and localized alcohol corneal epitheliectomy was done ahead of the mass. Triple freeze-thaw cryopexy was carried out to the scleral base and the edges of the conjunctiva. Fresh amniotic membrane was employed to cover the large conjunctival defect. The patient has been maintained on a close follow-up. **RESULTS:** No evidence of recurrence has been noted three years after the second extensive conjunctival resection, focal cryotherapy and use of amniotic membrane. Also, there is no clinical or radiological evidence of orbital or any distant metastasis.

CONCLUSION: Conservative surgical approach along with frequent follow-up and periodic systemic screening may allow successful local management of malignant fibrous histiocytoma, otherwise an aggressive conjunctival tumor.

AN UNUSUAL CASE OF RETROBULBAR ARACHNOID CYST AND ITS MANAGEMENT

Dipankar Das, Kasturi Bhattacharjee, Harsha Bhattacharjee, Jayanta Das, Ganesh Kuri, Sanjeev Handique

Sri Sankaradeva Nethralaya, Guwahati, India

PURPOSE: Arachnoid cysts are collections of cerebrospinal fluid contained inside a cavity lined by leptomeninges, constitute 1% of intracranial masses, mostly in the middle cranial fossa. Retro orbital arachnoid cyst in an elderly man is a rare occurrence. We report a rare case of retrobulbar arachnoid cyst in an elderly person with ophthalmic manifestations and its management. **METHOD:** A case report. A 73 years old man presented with poor vision in the left eye and headache for 3 weeks. He had mild proptosis, restriction of ocular movements with ill-sustained pupillary reaction in the left eye. **RESULTS:** Dilated fundoscopy of the left eye revealed disc edema and B-Scan USG showed a well-defined large cystic lesion in retrobulbar region compressing the optic nerve. CT-Scan revealed cystic lesion with watery density content in retrobulbar region with minimal proptosis. Under CT guided technique clear fluid was drained out and cytological and biochemical tests revealed arachnoid cyst. Immediately after cyst evacuation there was improvement of extra ocular movement. **CONCLUSION:** This fully documented case showed unusual presentation of retrobulbar cyst and its effective management.

A CYSTIC TUMOR OF THE ORBIT IN AN UNBORN BABY

Tero Kivela, Paivi Lindhal, Vilho Hiilesmaa

University of Helsinki, Helsinki, Finland

A 28-year-old woman, who was pregnant for the first time, underwent routine ultrasonography at 28 weeks of gestation. It was noticed that the fetus had a huge cystic tumor in her left orbit. Diagnosis and treatment will be presented as an "unknown".

EMBRYONAL RHABDOMYOSARCOMA, AN ANTENATAL DETECTED ORBITAL TUMOR

Rob De Keizer, Nicola, Marieke Sueters, Nicoline Schalijs-Delfos

LUMC, Leiden, The Netherlands

PURPOSE: Presentation of the first reported case of fetal orbital rhabdomyosarcoma in the third trimester of pregnancy. **METHODS:** Ultrasound examination at 34 + 3 weeks gestation revealed a large orbital tumor with ventrocranial displacement of the globe. **RESULTS:** A small for gestational age (SGA) girl was delivered by caesarian section at 37 + 6 weeks gestation. Postnatal MRI and biopsy led to the diagnosis of rhabdomyosarcoma. The tumor grew explosively and skin metastasis as well as sepsis developed at day 4. As curative doses of chemotherapy were considered too toxic, the child was not treated and died on day 5. On autopsy multiple metastasis in skin, liver, right lung, abdominal wall and left kidney were found. Histology showed a rhabdomyosarcoma with characteristics of the embryonal as well as alveolar type. **CONCLUSION:** Due to improved prenatal screening possibilities an orbital rhabdomyosarcoma was detected in a very early stage, which posed many unprecedented medico-ethical problems.

PRIMARY ORBITAL MELANOMA TREATED WITH IODINE 125 PLAQUE RADIOTHERAPY

Patrick dePotter, Lorette

Ocular Oncology Unit, Cliniques Universitaires St-Luc, Brussels, Belgium

A 58-year-old man reported left orbital discomfort for several weeks. Visual acuity was 20/20 OU. Anterior segment biomicroscopy and fundus examination were normal OU. Ultrasonography and orbital MRI demonstrated a left well-circumscribed retrobulbar mass. Incisional orbital biopsy via conjunctival approach confirmed the diagnosis of melanoma (epithelioid cell type). Systemic and dermatological evaluation revealed no evidence of primary melanoma elsewhere. The orbital tumor was managed with a non-shielded iodine 125 plaque. The patient developed radiation retinopathy 9 months after radiotherapy. Liver metastases were diagnosed after 20 month follow-up and successfully treated with chemotherapy. After 36 month follow-up, no orbital tumor recurrence or progressive metastatic disease was documented.

A CASE REPORT OF UNKNOWN ORBITAL TUMOR

Raman Mittal, Santosh Honavar, Geeta Vemuganti, Vijay Anand Reddy, Milind Naik, Ramesh Murthy,

LV Prasad Eye Institute, Hyderabad, India

A 28-year-old female presented with complaints of gradually increasing protrusion of the left eye for the past 4 months. There was no significant systemic or family history except for breast carcinoma in her elder sister. On examination, a firm, nodular, non-tender, immobile mass was present in the superotemporal orbit displacing the eyeball inferonasally. CT scan showed superotemporal orbital mass with bone erosion and intracranial extension. Incision biopsy of this unknown tumor was performed. Histopathology showed a malignant round cell tumor. Details of immunohistochemistry, final diagnosis, and further management will be discussed.

GIANT CHONDROID SYRINGOMA OF THE UPPER NASAL ORBITAL RIM: FIRST-REPORTED CASE

Ebrahim Shirzadeh, Alireza Ghasemi Aryan

Sabzevar School of Medical Sciences, Sabzevar, Iran

We describe the clinical manifestation, histology and differential diagnosis of a case of giant chondroid syringoma of the orbit. A 35-year-old man with a painless subcutaneous tumor of 3 x 3 x 4 cm in the upper nasal orbital rim was referred. The tumor was reported to have developed gradually over several years. The tumor was completely excised under general anesthesia. No recurrence was observed during 6-month follow-up. Macroscopically, a whitish, irregular and relatively soft mass with calcified spots in cut section was reported. Microscopically, tubules of various sizes were observed which were surrounded by cellular stroma and distinct areas of chondroid proliferation with no malignant changes. Chondroid syringoma may grow to a giant size as in our case. It may be considered in the differential diagnosis of a hard, slowly developing mass of the bony orbit. Despite the good prognosis, the recurrence of chondroid syringoma is reported when the tumor is not completely removed.

GLIOBLASTOMA OF THE OPTIC NERVE AND CHIASM

Thomas Cummings

Duke University Medical Center, Durham, NC, USA

Gliomas of the optic nerve and chiasm may be separable into two groups. Pilocytic astrocytomas tend to occur in patients younger than twenty years of age, have a favorable prognosis, and are associated with neurofibromatosis type 1. Malignant astrocytomas typically occur in adults and usually are fatal within one year. A 67-year-old female presented with complete loss of vision in her left eye and significant loss in her right eye. Contrast-enhanced T1 - weighted magnetic resonance images showed enlargement of the optic nerves and chiasm with contrast-enhancement of the orbital apices. The patient underwent a craniotomy for biopsy, which histologically showed cellular pleomorphism, mitotic activity, microvascular proliferation, and palisading necrosis typical of glioblastoma (WHO grade IV). She completed two weeks of whole brain irradiation and a post-operative MRI three months after biopsy showed tumor dissemination to the right occipital optic radiation and increase in size of the optic nerve and chiasm lesion. The patient expired five months following the craniotomy.

PLEOMORPHIC ADENOMA OF LACRIMAL GLAND IN A 7-YEAR-OLD BOY: A CASE REPORT

Vikas Mittal, SL Adile, Ashok Chandrakar, Mange Garg, Ruchi Juneja, Anil Gupta

Pt. JNM Medical College, Raipur, India

Pleomorphic adenoma is the most common epithelial tumor of lacrimal gland presenting typically in middle age. Very few cases of pleomorphic adenoma of lacrimal gland in pediatric age group have been reported in world literature. We report a 7-year-old Indian boy having pleomorphic adenoma of right lacrimal gland with photographic documentation. 7 years old male child presented to us with a 3-year history of painless, firm lacrimal gland mass measuring 3cms x 2cms with a small part of it prolapsing into superior fornix. There was 5 mm proptosis with inferonasal displacement of right eyeball. CT scan revealed a well defined 2.5cms x 3.2cms mass arising from right lacrimal gland causing enlargement of lacrimal fossa without any bony erosion. Lateral orbitotomy was performed under general anesthesia and the mass was excised intact. Histopathological examination demonstrated ductules of benign epithelial cells and proliferation of myoepithelial cells in a myxoid stroma consistent with pleomorphic adenoma. Although rare, pleomorphic adenoma of lacrimal gland should be considered in the differential diagnosis of pediatric orbital masses because of prognostic importance of removing the tumor intact.

BASAL CELL ADENOCARCINOMA OF LACRIMAL GLAND: A NEWLY RECOGNIZED RARE TUMOR

Subhash Betharia, Seema Kashyap, Vidushi Sharma, Seema Sen, Harish Pathak, Vijay Wagh

Rajendra Prasad Center for Ophthalmic Sciences, New Delhi, India

Basal cell adenocarcinomas have recently been included as a differential diagnosis in the tumors of salivary glands. The first such case in a lacrimal gland was reported in 2000 and there have been extremely few reports on this entity since then. We report the case of a 40 year old man, who presented with proptosis due to a lacrimal gland mass. There was no history of pain or diplopia. On computed tomography (CT) scan, a mass was seen arising from the lacrimal gland, with unusually extensive calcification. There was no obvious bony erosion and the mass crossed the vertical orbital midline. A lateral orbitotomy was performed and the mass was removed in toto. Histopathological examination showed features diagnostic of basal cell adenocarcinoma, including extensive

squamous metaplasia. In view of this diagnosis, the patient was advised serial follow-up. It is important to differentiate this tumor from the much more aggressive and fatal adenoid cystic carcinoma, specially its basaloid variant, as basal cell adenocarcinoma carries a much more favorable prognosis.

PRIMARY DUCTAL ADENOCARCINOMA OF THE LACRIMAL GLAND: A RARE ENTITY

Harish Pathak, Subhash Betharia, Seema Sen, Seema Kashyap, Vijay Wagh, Vidushi Sharma

Dr. Rajendra Prasad Center for Ophthalmic Sciences, New Delhi, India

A 47-year-old man presented with gradual protrusion of right eye of three months duration associated with vision loss. On examination there was severe proptosis with inferonasal displacement of the right eye. CT scan of orbit revealed a non-encapsulated heterogeneous mass lesion superotemporally extending into the intraconal space abutting the optic nerve. A provisional clinical diagnosis of lacrimal gland carcinoma was made and a wedge biopsy was performed. On light microscopy a high-grade adenocarcinoma with extensive desmoplastic reaction, necrosis and calcification was seen. Microscopically both intraductal and infiltrating duct components along with perineural infiltration was present. Systemic evaluation was done with no evidence of a primary or secondary tumor in the body. A final diagnosis of primary ductal adenocarcinoma (PDA) of the lacrimal gland was made. Exenteration of the right orbit was done. Histopathology confirmed the diagnosis. Patient received adjunctive radiotherapy and showed no evidence of recurrence on subsequent follow-up. PDA of the lacrimal gland is a rare entity and is a histological equivalent of salivary duct adenocarcinoma. To the best of our knowledge this is the first case of ductal adenocarcinoma of the lacrimal gland in India and the third case worldwide.

SOLITARY PLASMOCYTOMA OF ORBIT

Nagendra Shah, Usha Kim

Aravind Eye Hospital, Madurai, India

A 70-year-old systemically healthy man presented with progressive gradual diminution of vision with protrusion of the left eye for six months. He had 7 mm axial proptosis with ptosis and restricted ocular motility. Vision in the left eye was reduced to hand motion and there was relative afferent pupillary defect. CT scan showed an orbital mass extending up to the apex compressing the optic nerve, and into the ethmoid sinus. There was scalloping of the medial orbital wall. Systemic investigations were unremarkable. Initial biopsy was suggestive of neurofibroma. A repeat biopsy showed monomorphic plasma cells with lobulated, eccentric and basophilic nuclei with cartwheel arrangement of chromatin suggestive of plasmacytoma. There was no systemic plasmacytoma. External beam radiotherapy resulted in local tumor regression. Solitary orbital plasmacytoma is rare.

BILATERAL ORBITAL LYMPHOMA: A RARE PRESENTATION OF MULTICENTRIC SYNCHRONOUS EXTRANODAL MALT LYMPHOMA

Gaurav Bahl, Supriya Chopra, S Ramani, Siddharth Laskar, K Naresh, M Muckaden

Tata Memorial Hospital, Mumbai, India

Primary orbital lymphomas form a very small subgroup of lymphomas. Bilateral multicentric extranodal lymphomas are even rarer. We report a case of bilateral orbital lymphoma associated with preauricular nodal enlargement along with

bilateral breast lymphomas. A 50-year-old lady presented with bilateral orbital swelling since the last 12 years. This was associated with bilateral preauricular nodal enlargement. On systemic examination there were associated bilateral breast lumps. Biopsy was suggestive of MALT Lymphoma. On detailed clinico-radiological evaluation there was no evidence of systemic involvement. Review of published literature on this subject revealed this to be the first reported case of bilateral synchronous extranodal MALT lymphoma involving 3 sites. As this clinical presentation is very rare, there is paucity of guidelines regarding the management of this particular entity. Various case series have analyzed the outcome of orbital and breast lymphomas occurring independently. Primary extranodal MALT lymphomas are indolent in nature and long-term loco-regional control and survival is seen even with single modality treatment. However due to multisite involvement combined modality treatment was offered to this patient. Details of the same shall be discussed.

BILATERAL ORBITAL NON-HODGKIN'S LYMPHOMA IN A PATIENT WITH ACQUIRED IMMUNODEFICIENCY SYNDROME – A CASE REPORT AND REVIEW OF LITERATURE

Mallipatna C Ashwin, Gosala RK Sarma, Ravindra R Battu, Ringhoo T Jose

St. John's Medical College Hospital, Bangalore, India

We report an unusual presentation of orbital Non-Hodgkin's Lymphoma (NHL) associated with HIV and discuss the review of literature pertinent to orbital and ocular NHL. A 30 year old male with AIDS presented with complaints of headache and nasal stuffiness, proptosis of the left eye of one month duration, and associated with eye pain and progressive loss of vision in both eyes of fifteen days duration. Examination revealed a non-tender abdominal mass in the right lumbar region and a facial swelling over the paranasal sinuses. On ophthalmologic examination the visual acuity was reduced to finger counting in the right eye and no perception of light in the left eye. The patient had a proptosis of the left eye with total ophthalmoplegia and a right partial ophthalmoplegia. MRI showed a diffuse sino-nasal mass lesion infiltrating the orbits, orbital apices, parasellar region, left cavernous sinus and retropharyngeal region. Functional endoscopic sinus surgery with clearance of pus was done and sinus tissue from right maxillary sinus was sent for histopathology. This showed an infiltrate suggestive of NHL. Immunohistochemistry showed CD 20 positivity consistent with a B-cell NHL.

SCLEROSING EXTRAMEDULLARY HEMATOPOIETIC TUMOR IN THE ORBIT

Lai Yuen, Kei Kwok, Nongnart Chan

Hong Kong Eye Hospital, The Chinese University of Hong Kong, Hong Kong, China

A 62-year-old with a known history of myelofibrosis presented with a 3 x 2 cm left lower lid mass in the anterior orbit. CT scan showed bilateral multiple intraconal and extraconal orbital soft tissue densities. A multilobulated rubbery pinkish tumor adhering to surrounding tissue was identified during a left anterior orbitotomy. Histopathological examination disclosed a lesion composed of whorls of myxoid to sclerotic stroma with thick collagen strands. Large and atypical megakaryocytes, granulocytic and erythroid precursors were scattered throughout the stroma. Immunohistochemical staining showed positive result for myeloperoxidase, confirming presence of granulocytes. Some of the megakaryocytes showed faint staining for factor VIII related antigen. No atypical lymphoid or myeloid blast population was present. The diagnosis of sclerosing extramedullary hematopoietic tumor was made. Her vision remained stable 3 years after presentation. Orbital sclerosing extramedullary hematopoietic tumor is a rare condition. Although the imaging appearances look striking, this condition runs a relatively benign course and aggressive surgery seems not necessary.