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Dr. TATYANA MILMAN declares she has no conflicts of interest to disclose.

The Bony Orbit
7 Bones
The Roof

Frontal Bone

The Medial Wall

The Inferior Wall

The Lateral Wall

And

Palatine Bone
(not shown here)
Primary lesions of the bony orbit

0.6%-2% of all orbital tumors

Primary osseous and fibro-osseous lesions:
~40%

Primary osseous and fibro-osseous orbital lesions

Benign orbital fibro-osseous lesions
- Fibrous dysplasia
- Ossifying fibroma
- Osteoma
- Osteoid osteoma and osteoblastoma

Interesting cases

Fibrous dysplasia

Definition
- Dysplastic skeletal anomaly
- Distortion of normal medullary bone and replacement with immature woven bone

Clinical presentation
- First 2 decades (most patients <30)
- M = F

Monostotic fibrous dysplasia

Fibrous dysplasia

Clinical presentation
- First 2 decades (most patients <30)
- M = F

Polystotic fibrous dysplasia in McCune Albright Syndrome
http://tumorlibrary.com/
Fibrous dysplasia

Clinical presentation
- First 2 decades (most patients <30)
- M = F

Craniofacial dysplasia

Orbital wall involvement

Pathophysiology / Genetics
- GTPase mutations
  - Codon 201 Exon 8
  - ~60%

CREB pathway
**GNAS and fibrous dysplasia**

**Fibrous dysplasia**

**Imaging**

[References](http://radiopaedia.org/images/)

[References](http://reference.medscape.com/)

**Low-grade central osteosarcoma**

Amplification of 12q13-15 (MDM2 and CDK4 genes)

MDM2 and CDK4 protein overexpression
Fibrous dysplasia

Diagnosis
- Clinical – radiographic – pathologic correlation

Prognosis
- Typically quiescent after puberty
- Occasionally persistent growth
- Malignant transformation
  - Osteosarcoma > chondrosarcoma / fibrosarcoma

Management
- Observation
- Bone contouring
- Curettage / resection
- Optic nerve decompression / radical resection
- Bisphosphonates
- Radiation contraindicated

Ossifying fibroma

Definition
- Benign bone producing neoplasm, composed of fibrocellular tissue and mineralized material of varying appearances

- Craniofacial skeleton – 2 variants:
  1) Ossifying fibroma of odontogenic origin (cemento-ossifying fibroma, ossifying fibroma NOS)
  2) Juvenile ossifying fibroma
     - Psammomatoid variant
     - Trabecular variant
Ossifying fibroma

Pathophysiology / Genetics

No GNAS1 mutations

Orbital ossifying fibroma:
• Non-random chromosome break points at Xq26 and 2q33 - t(X;2)

Psammomatoid ossifying fibroma
• MDM2 gene amplifications WITHOUT protein overexpression


Clinical presentation

• Average at presentation 16-33 years (range 3 mo-72 yrs)
• F > M


Imaging

Ossifying fibroma

Diagnosis
- Clinical – radiographic – pathologic correlation

Management and Prognosis
- Progressive growth
- Multiple recurrences following incomplete excision
- Complete excision recommended
- Malignant transformation not reported

Osteoma

Definition
- Benign lesion composed of mature bone with predominantly lamellar structure
- Almost exclusively identified in craniofacial skeleton

Pathophysiology / Genetics
- Traumatic, infectious and developmental theories
- No well-characterized genetic alterations

Association with Gardner syndrome
Osteoma

Clinical presentation
- 4th – 5th decades (10 - 82 yrs)
- M = F vs. M:F = 2:1
- Most are asymptomatic and incidental

http://www.sarawakeyecare.com/

Osteoma

Imaging

http://www.archivesofpathology.org/

Ivory osteoma
Osteoma

Management and Prognosis

- Observation if asymptomatic
- Complete surgical resection (curative):
  - Symptomatic
  - Sphenoid sinus lesions

Osteoid osteoma and osteoblastoma

Definition
- Benign osteoblastic tumors with overlapping clinical, radiographic and histologic findings

Osteoid osteoma:
- <1.5 cm in greatest dimension
- Extremely rare in the head and neck region

Osteoblastoma:
- >1.5 cm in greatest dimensions
Osteoid osteoma and osteoblastoma

Pathophysiology / Genetics

- Not well understood
- Potential genetic overlap (clonal chromosomal abnormalities)
- **Osteoid osteoma**
  - Distinct structural chromosomal alterations (22q13)
- **Osteoblastoma**
  - Three-way translocations involving Chr 1, 2, 14
  - Rearrangement of 1q42

Clinical presentation

- Distinct predilection for males, 10 – 20 years
- Nocturnal pain in extraorbital lesions

Clinical presentation

- Distinct predilection for males, 10 – 20 years
- Nocturnal pain in extraorbital lesions

**Imaging**

- Sclerotic rim
- Nidus

**Pathology**

- Rim of sclerotic reactive lamellar bone
- Central nidus
Epithelioid (aggressive) osteoblastoma

Osteoblastoma-like osteosarcoma

- MDM2, CDK4, TOP2A, MACC1 amplification
- Complex chromosomal alterations
- LOH for Chr 3q, 13q, 17p, 18q
- TP53 and RB1 mutations
Osteoid osteoma and osteoblastoma

Management and Prognosis

Osteoid osteoma
- Conservative surgical resection curative

Osteoblastoma
- Can be locally aggressive (epithelioid osteoblastoma)
- Recurrences following incomplete resection or piecemeal removal
- Complete excision recommended
- Risk of malignant transformation into osteosarcoma??
  • Debated

Selected cases

Osseous Tumor of the Orbital Bone
Nasreen A. Syed, M.D.
P.C. Boni Eye Pathology Laboratory
University of Iowa

Fibro-osseous orbital lesion
Michele M. Bloomer, MD
Department of Ophthalmology
UCSF

Midfacial Mass
Sander R. Dubovy, MD
Florida Lions Ocular Pathology Laboratory
Bascom Palmer Eye Institute
University of Miami Miller School of Medicine

15 y.o. Haitian girl with 3 year history of growing right mid-facial mass
Ossifying fibroma
Treatment Goals

• En-block resection of tumor

• Reconstruction:
  • Floor of orbit
  • Right maxilla
  • Function
  • Cosmesis
8 year old boy, previously healthy
Progressive right proptosis x 6 weeks

2.7 x 1.8 x 1.6 cm mass

Anterior orbitotomy with piece-meal removal of the lesion
Diagnosis?

Diagnosis rendered in consultation with bone pathologist:

“Benign osseous neoplasm most consistent with osteoid osteoma”
Diagnosis

Diagnosis rendered in consultation with outside bone pathologist:

"Osteoblastic variant of osteosarcoma"
Fibro-osseous orbital lesion

Michele M. Bloomer, MD
Department of Ophthalmology
University of California, San Francisco

Verhoeff Zimmerman Society Meeting
April 29 – May 2, 2010
Hyatt Regency, Sarasota

65 year old, otherwise healthy Asian man with right eye swelling and diplopia x 6 weeks
Disease Course

- Immediate post-operative image revealed near gross total resection
- No alteration in the residual mass >1 year after resection
Paget’s Disease

- Most patients >55 years
- Skull frequently affected
- Presents with diplopia and globe displacement
- Polyostotic and monostotic forms
- Radiographically similar to fibrous dysplasia
- Can be self-limited
- Rare in Asians

Paget’s Disease

- Disordered bony remodeling
  - Early: osteoblastic and osteoclastic rimming
  - Late: burnt out phase
- Classic histologic feature “jigsaw puzzle”
- Occasional transformation into osteosarcoma

Primary osseous and fibro-osseous orbital lesions

- Benign tumors
  - Fibrous dysplasia
  - Ossifying fibroma
  - Osteoma
  - Osteoblastoma

- Challenges in pathologic diagnosis
  - Fragmented nature of the specimens
  - Significant clinical, histologic, and radiographic overlap

- Need for careful clinical-radiographic-pathologic correlation

PLEASE TURN OFF YOUR CELL PHONES
THANK YOU