CASE 7

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Case History

• A 66 year-old man

• Exophthalmos of the left eye

• Normal visual acuity
Orbital MRI

- Increased orbital fat extending through the conjunctival space in the supero-temporal quadrant
Orbital MRI

- Increased orbital fat extending through the conjunctival space in the supero-temporal quadrant
An excision was performed

- 2.5 cm fragment of normal adipose tissue
Diagnosis:
SUBCONJUNCTIVAL HERNIATED ORBITAL FAT (HOF)
Orbital Fat : 2 components

- **Intraconal:**
  - Limited by the extraocular muscles and the Tenon capsule (*fascia bulbi*)

- **Extraconal:**
  - Limited posteriorly by the peristium and the extraocular muscles
  - Limited anteriorly by the orbital septum

*Schmack I. Am J Surg Pathol 2007*
Herniation of orbital fat tissue

- Intraconal herniation of fat tissue:
  - Frequent, under-reported
  - Obese patients
  - Men > Women
  - 28-94 y (mean age 65 y)
  - Younger patients: after trauma/surgery
Subconjunctival herniated orbital fat: clinical aspect

• Supero-temporal fornices

• Bilateral (+++), asymmetric

Kantelip B. AIP N°60
Subconjunctival herniated orbital fat: clinical aspect

- Less frequently: inferotemporal quadrant

Stacy RC. Histopathol 2014.
Shields JA, Shields C. Eyelid, Conjunctival and Orbital Tumor
Subconjunctival herniated orbital fat: clinical aspect

- Yellow-to orange
- Elevated, convex-shaped, compressible
- Thin vessels on the surface
- DD: lacrymal lesions

Courtesy of Dr Flausse
Imaging findings

- Fat tissue extending from the intraconal space into the subconjunctival area
Microscopic findings

• Non encapsulated fat tissue
• Mature adipocytes
• Thin septae
Floret cells

- Multinucleated cells
- Small hyperchromatic, uniform nuclei
- Within the septae, not in the lobules
- May be numerous
- Reactive cells?
Lochkern cells

- Pathognomonic
- Vacuolated nuclei
- No atypia, no mitoses
- No lipoblasts
In routine practice, only MDM2 is useful but not obligatory.
Immunohistochemistry : RB

- Main DD: pleomorphic lipoma
- Only 1 study published
- No loss of expression in 5 tested cases

Molecular findings: aCGH

Stacy RC. Histopathology, 2014.
Follow-up

- No recurrence
- No malignant transformation
Differential diagnoses

• Pleomorphic lipoma
• Atypical lipomatous tumor / well differentiated liposarcoma
• Dermolipoma
• Other liposarcomas
Orbital pleomorphic lipoma

• Floret cells:
  – In the lobules more than in the septae
  – Large hyperchromatic nuclei

• Immunohistochemistry : Loss of RB

• Rarely reported in this location:
  – Past cases were mostly bilateral*
  – Does it truly exist?

Pleomorphic lipoma

HOF

Courtesy of Dr Neuville (Strasbourg)

Present case
Atypical adipocytic tumor/Well differenciated liposarcoma

• Rarely reported in the orbit

• Past cases: some were bilateral (HOF???)

• Recent cases:
  – Typical morphology
  – Typical MDM2 positivity
  – Typical MDM2 amplification (FISH)
Atypical adipocytic tumor/Well differentiated liposarcoma

Subramamian MM. Histopathology, 2015.
Dermolipoma

- Congenital, but diagnosed in young adults
- Superotemporal, extending into the entire external fornix
- Unilateral
- May be associated with Goldenhar syndrome
- Covered by acanthotic conjunctival epithelium
- Connective tissue with abundant adipose tissue

Shields JA, Shields C. Eyelid, Conjunctival and Orbital Tumor
Orbital liposarcomas

• Quite rare
• Epidemiological data is controversial: Female or male predominance?
• Mean onset age: 40-45 years
• Exophthalmos, diplopia, pain, optic nerve compression
• Subtypes: Myxoid liposarcomas (60%) > ALT (30%) > Pleomorphic liposarcomas (10%)
• No histological aspect specific to the orbit compared to other locations
• Treatment: exenteration, adjuvant radiation or chemotherapy is debated

Bordolla-Pertiera AM. Arch Soc Esp Oftalmol. 2017
Take home messages

• Herniated subconjunctival fat tissue: benign, under-reported
• Elderly, obese men
• Superotemporal quadrant
• Mature adipose tissue
• Floret cells in the septae
• Lochkern cells in the lobules
• MDM2: negative
• DD : Atypical lipomatous tumor
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