Disclosure of Relevant Financial Relationships

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Bone and soft tissue eve conference- F Le Loarer

Disclosure of Relevant Financial Relationships

Dr. Francois LE LOARER has no conflict(s) of interest or NON academic affiliation to declare

CLINICAL PRESENTATION

• 15 yo boy
• No medical history
• Isolated 1cm pimpl in the back
• Not pigmented
• Known for at least 1 year
  >Excision biopsy
Mitotic index: 25/10HPF

FIRST ROUND IMMUNO PANEL

AE1/E3

EMA

FIRST ROUND IMMUNO PANEL

CD99

S100

CD34, DESMIN, MYOGENIN: NEGATIVE

DIFFERENTIAL: MELANOMA

• Junctional component
DIFFERENTIAL: RHABDOID TUMORS
- Infants <3yo++
- Round cell pattern
- High grade
- Keratin expression

DIFFERENTIAL: ROUND CELL SARCOMA
- "The big three"
  - Ewing Sarcoma
  - CIC-rearranged Sarcoma
  - BCOR-rearranged Sarcoma

SECOND ROUND IMMUNO PANEL
- SOX10
- HMB45 & MELAN A

1-EWING SARCOMA
- CD99
- EWSR1/FUS fusions with different partners FLI1, ERG...
**2-CIC-SARCOMA**

- 

**CIC-SARCOMA**

- 

**3-BCOR-SARCOMA**

- 

**SECOND ROUND IMMUNO PANEL**

- 

**DIFFERENTIAL: MYOEPITHELIAL TUMOR**

- Intermediate malignancy
- Myoepithelial differentiation

- 

**EWSR1 rearrangements in half of cases**
MALIGNANCY IN MYOEPITHELIAL TUMOR

• "Myoepithelial carcinoma"
• Criteria in soft tissue: cytologic atypia!!
• Recurrence and mets rates: 40%
• Loss of SMARCB1
• More frequent in pediatric patients: 50%

SECOND ROUND IMMUNO PANEL

P63
SOX10

AML, GFAP: negative
SMARCB1: retained

MOLECULAR TESTING

• Break-apart FISH EWSR1 probe

PHONE CALL PEDIATRICIAN:
"WHAT's Ur Diagnosis?"

• "Myoepithelial" phenotype
• EWSR1 rearrangement

ATYPICAL MYOEPITHELIAL TUMOR

-No Grading (no prognostic value)
-Warnsme but not fitting myoepithelial carcinoma

EWSR1:
a "TRANSLOCATION HUB"


RT-PCR Molecular report:
EWSR1: a "TRANSLOCATION HUB"
« SUPERFICIAL » EWING SARCOMA

- Same features than deep-seated ES:
  - age, morphology, location, molecular
  - Local presentation
  - Unique nodule present for 1-15 months…
  - Size 6-100mm
  - Better prognosis than deep-seated ES?
  - Adverse outcome when local treatment (Mets and DOD)

CLINICAL FOLLOW UP

- Body TDM: normal
- Stage 2 disease (T1A, N0, M0, High grade)
- Ewing type Chemotherapy regimen
- No adverse event at 8 mo follow-up

TAKE HOME MESSAGES

- ES express Keratins in 30%…
- Coexpression of Keratins and S100 not specific myoepithelial lineage
- EWSR1 is a « translocation hub »
- Don't try to be nice to clinicians…

BEWARE OF MYOEPITHELIOMA…

BONE & SOFT TISSUE EVE SPECIALTY CONFERENCE

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