CASE 6

Milanka Živanović
On behalf of Boštjan Luzar, MD, PhD

Institute of Pathology
Medical Faculty University of Ljubljana
SLOVENIA
CASE 6

- 62-year-old male
- Nodular lesion on the sole of the foot of unknown duration
- Clinical diagnosis: pyogenic granuloma
Penny
for your thoughts
### IMMUNOHISTOCHEMISTRY

<table>
<thead>
<tr>
<th>Marker</th>
<th>Result</th>
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</thead>
<tbody>
<tr>
<td>CD99</td>
<td>+++</td>
</tr>
<tr>
<td>S100</td>
<td>focally +</td>
</tr>
<tr>
<td>SYNAPTHOESISYN</td>
<td>focally +</td>
</tr>
<tr>
<td>Melan A</td>
<td>—</td>
</tr>
<tr>
<td>HMB45</td>
<td>—</td>
</tr>
<tr>
<td>Tyrosinase</td>
<td>—</td>
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<tr>
<td>CK-MNF116</td>
<td>—</td>
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<tr>
<td>Chromogranin</td>
<td>—</td>
</tr>
<tr>
<td>Desmin</td>
<td>—</td>
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<tr>
<td>LCA</td>
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MOLECULAR TESTING
(flourescent in situ hybridisation)

Translocation t(11;22)
1° CUTANEOUS EWING’S SARCOMA / PERIPHERAL PRIMITIVE NEUROECTODERMAL TUMOUR
Ewing’s sarcoma of the bone ("diffuse endothelioma of bone" James Ewing, 1921)
Primary Cutaneous Ewing’s Sarcoma / PPNET

- Rare malignant neoplasm (less than 100 cases reported)
- Female predominance (2:1)
- Median age at diagnosis is between 2nd and 3rd decade of life (broad age distribution: 2 to 67 years)
- CLINICAL PRESENTATION -

- Well defined nodule
- Median tumour size 2-3 cm (range from 0,5 to 12 cm)
- Predilection for lower and upper limbs, head and trunk
- Usually asymptomatic, but can be painful
- Gross appearance – pink to brown, soft, hemorrhagic
PRIMARY CUTANEOUS EWING’S SARCOMA / PPNET

HISTOLOGY

- Non-encapsulated, poorly circumscribed tumor located in dermis and/or subcutis
- Pattern of growth: solid sheets, lobules, trabecules
- Composed of small to medium sized cells with round pleomorphic nuclei and scant eosinophilic cytoplasm
- Pseudocistic spaces
- Haemorrhage
- Necrosis
- Mitotic activity (median from 6 to 8 per 10 high-power fields)
Diffuse, strong positivity for CD99

Variable, patchy positivity for S100, various cytokeratins (up to 30% of cases), neuroendocrine markers (NSE, chromogranin, synaptophysin), GFAP, NKI-C3, neurofilament protein, myogenin and desmin

Negative for HMB45, Melan A, EMA, CD45, CD3, CD20, CD43, myeloperoxidase
Translocation t(11;22): fusion of EWS gene on chromosome 22 to FLI gene on chromosome 11 (90% of cases)

Translocations pairing EWS gene with other members of ETS family of genes, namely ERG gene at 21q22, ETV1 gene at 7p22 and FEV gene at 2q33 (10% of cases)
Primary Cutaneous Ewing Sarcoma
A Systematic Review Focused on Treatment and Outcome

M. Delaplace, C. Lhommet, G. de Pinieux, B. Vergier, A. de Muret, L. Machet

2012

• more indolent clinical course than their bone or deep soft tissue counterparts (smaller tumours, earlier diagnosis… !?)
All EFT are treated in the same way:

1. surgical excision
2. radiotherapy (in case of incomplete surgical removal)
3. combined chemotherapy (Euro-Ewing 99 protocol)
In children and young adults

- Rhabdomyosarcoma
- Neuroblastoma
- Metastatic Ewing’s sarcoma/PNET
- Lymphoblastic lymphoma
In adult patients

- Merkel cell carcinoma
- Metastatic neuroendocrine carcinoma (pulmonary and extrapulmonary sites)
- Poorly differentiated adnexal tumours
- Melanoma
- Clear cell sarcoma
DEATH BY POWERPOINT