Special slide seminar

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Case history

- ♀, 33 years old

- resistance on the lateral edge of the left knee-cap with progressive pain developing within the last six months

- no history of injury or overload
CT scan

- mass (57x45x27mm) lateral to the patella, containing calcifications
- same density as the patellar cartilage
- not connected to the bone
- two other lesions (6x12mm and 25x22mm) in the popliteal fossa (suspicious for metastatic lymphadenopathy)
macro

- well-demarcated, partially nodular 60x40x25mm tumor with whitish color
micro... Gömöri
micro... HE
micro... HE
micro... HE
micro... HE
micro... HE
ihc – S-100
ihc – HMB-45
• negative:
  – melan A
  – CD117 (c-kit)
  – CK (AE1/AE3)
  – EMA
  – SMA
  – desmin
  – myo-D1
  – CD 57
ihc – Ki-67
glycogen
ultrastructually
Clear cell sarcoma of soft parts
(malignant melanoma of soft parts)
discussion

- rare malignant tumor of soft tissue
- Enzinger 1965
- young adults (20 - 40 y. o.)
- deeply located tumor most often of the extremities (40% foot and knee region),
- only exceptionally head, neck or chest
discussion

• clinically: slow-growing, sometimes painful mass in the deep soft tissue without skin involvement

• macro: lobulated to nodular tumor, white-gray color attached to tendon or aponeurosis, size 2-6 cm, +/- hemorrhage, necrosis, cystic changes, occasionally focal brown pigmentation
micro:

- Nests and trabeculae of spindle to round cells separated by fibrous septa merging to adjacent tendon or aponeurosis
- typical cells: vesicular nuclei, prominent basophilic nucleoli, abundant clear or pale eosinophilic cytoplasm containing glycogen
- +/- multinucleated giant cells
- Atypia only exceptionally, mitosis not common (X recurrence or metastasis)
- melanin not present or only in small amount (Fontana)
discussion

- IHC:
  + S-100 protein, HMB-45 (80%)
  + melan-A (40%)
  + MiTF (70%)
  - actin, desmin, CD117 (c-kit)

- ultrastructurally : melanosomes or premelanosomes in cytoplasm
• Tumor of neuroectodermal origin with melanocytic differentiation (typical micro, ihc, ultrastr.)
• 1983 Chung a Enzinger: malignant melanoma of soft parts
• chromosomal translocation t(12;22)(q13;q12) => separate entity
• prognosis unfavorable, high-grade sarcoma (slow but continuous progression)
• local recurrence and metastasis (LN, distant)
• no histological grading
• worse prognosis: size > 5 cm, necrosis, distant metastasis
• Th.: – wide surgical excision
  – chemo and radioresistant
1. melanocytic tumors with positive HMB 45:

- **MM (nodular form)** or metastasis of MM: primary tumor usually in skin, frequent atypia and mitosis, in 65% CD117+, different alteration of DNA (no t(12;22)(q13;q12) translocation)

- **cellular blue nevus**: dermis (back, LE), inconspicuous nucleoli, no mitosis and necrosis

- **malignant blue nevus**: typically head and legs, superficial localization, like cellular blue nevus with atypia, mitosis (+ atypical) and necrosis

- **PEComa**: smooth muscle + melanocytic differentiation, HMB-45 +, melan-A+, SMA+ (v 90%), desmin + (35%), S100 protein only 30%

- **paraganglioma-like dermal melanocytic tumor**: dermis, inconspicuous nucleoli, no necrosis
2. spindle cell tumors:
- **fibrosarcoma**: vimentin +, HMB-45 -
- **epitheloid leiomyosarcoma**: actin+, desmin+, HMB-45 -
- **MPNST (malignant schwannoma)**: associated with nerves, neurofibromas or Recklinghausen d., pleomorphic spindle cells with dark nuclei and clear cytoplasm, high mitotic activity, rare positivity of melan-A and HMB-45
- **monophasic synovial sarcoma**: hypercellular with uniform pattern, dark bland nuclei with coarse chromatin, frequent calcification, hyalinization and osseous metaplasia, mast cells, CK + (50-80%), negative melan-A and HMB-45
- **paraganglioma**: chromatin „salt and pepper“, chromogranin +, synaptophysin +, S-100 protein positive sustentacular cell