Leiomyosarcoma

Leiomyosarcoma is a malignant tumor derived from Smooth muscle cells. Leiomyosarcomas are relatively rare tumors, most often occurring in the limbs, Retroperitoneum, gastrointestinal tract and female genital tract. The age-standardized incidence is around 9 cases per million. The incidence is higher in women, after exclusion of uterine leiomyosarcomas, the incidence is comparable. Leiomyosarcomas are tumors of older age, they are quite rare before the age of 20, and the incidence increases with age. The overall five-year survival is around 50%, a slightly upward trend is evident. Overall survival varies according to the location of the tumor, while in uterine leiomyosarcomas the five-year survival is around 40%, in the case of skin leiomyosarcomas the five-year survival is around 95%.

Clinical picture

Clinically, the tumor manifests itself primarily as a massive mass. In addition to pain, depending on the location, it can present itself, for example, as Budd-Chiari syndrome, renal failure due to renal vein compression, or abnormal vaginal bleeding.

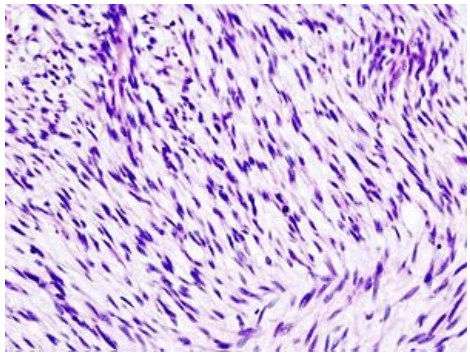
The tumor spreads per continuity, grows into blood vessels and establishes distant hematogenous metastases, especially in the lungs.

Macroscopic behavior

The tumor impresses macroscopically as a mass, the color can be gray, white or brownish to pink. The structure can be fascicular as well as soft and homogeneous. Macroscopically, it can be both relatively well-defined and infiltratively growing into the surroundings. The incision may be relatively homogeneous, especially in larger tumors, secondary regressive changes, ie hemorrhage, necrosis and cyst formation, are common.

Histopathology

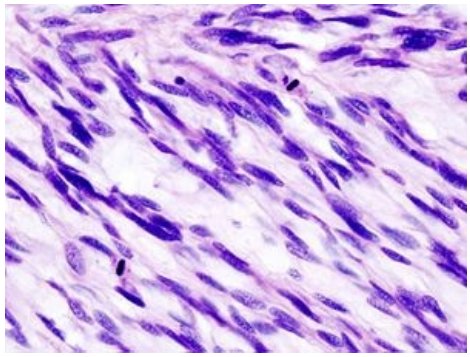
An intersecting well-defined groups of spindle cells are a typical histopathological pattern . However, this pattern is not fully established and the tumor may acquire a storiform, palisade-like or hemangiopericytoma-like pattern in some areas. The tumor is usually highly cellular, but fibrosis or myxoid changes can occur (myxoid leiomyosarcoma). Myxoid changes can be so marked that the tumor is reticulate or microcystic in nature. Hyalinization and necrosis zones are common in large tumors. Rather, exceptionally, the tumor may be inflammatory infiltrated.



Uterine leiomyosarcoma, H&E. The characteristic arrangement of the cells is evident.

Tumor cell nuclei are usually elongated with blunt ends, they may be lobed or serrated. Nuclei are usually hyperchromatic, nuclear pleomorphism is noticeable, but sometimes it can be only mild or noticeable only in deposits. Mitotic activity is pronounced, atypical mitoses are common. The cytoplasm is eosinophilic to clear, intracytoplasmic vacuoles are relatively common.

Epithelioid leiomyosarcoma is an uncommon variant in which osteoclast-like huge multinucleated tumor cells are present surrounded by marked chronic inflammatory infiltration. Often this variant is only visible in foci.



Uterine leiomyosarcoma, H&E. The characteristic blunt rounding of the ends of the elongated cores is evident.

Immunohistochemical properties

Practically always positive:

- SMA,
- desmin,
- h-caldesmon,
- histon-deacetylase 8.

It can be positive:

- keratin,
- EMA,
- CD34,
- S100,
- CD10.

Usually negative:

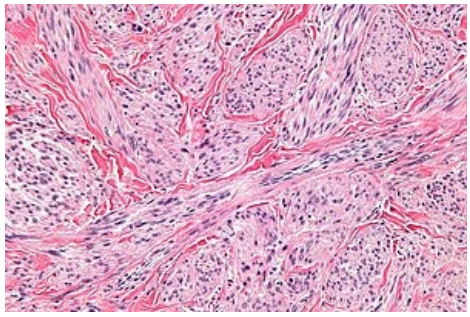
- KIT (in the positive case no c-Kit gene mutation is present).

May be positive in uterine leiomyosarcomas:

- estrogen receptors,
- progesterone receptors,
- androgen receptors.

General markers:

- Ki67 shows higher activity in leiomyosarcomas compared to leiomyomas.
- P53 is mutated in about a quarter to a half of uterine leiomyosarcomas (and is never mutated in leiomyomas).


Cutaneous leiomyosarcoma, H&E

Differential diagnostics

It is necessary to differentiate leiomyosarcoma differentially diagnostically, especially from leiomyoma and other smooth muscle proliferations, especially atypical proliferation. At the border between leiomyomas and leiomyosarcomas, there is a lesion of uncertain biological behavior called STUMP (Smooth Muscle Tumor of Uncertain Malignant Potential).

Link

Visual preparations

Template:Visual preparation

related articles

- Leiomyoma
- Mesenchymal tumors
- Malignant tumors of the uterine body

Reference

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External link

- de.Pathowiki. *Leiomyosarkom* [online]. [cit. 3/2014]. <http://de.pathowiki.org/wiki/Leiomyosarkom Leiomyosarkom>.

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