Classification of Solitary Fibrous Tumors (SFTs)

Solitary fibrous tumors (SFTs) are rare mesenchymal neoplasms that can occur in any soft tissue, including the meninges and spinal cord. Their classification is primarily based on histopathological and molecular characteristics, as outlined in the **WHO Classification of Tumors of Soft Tissue and Bone (2020)** and the **WHO Classification of Tumors of the Central Nervous System (2016)**.

Histological Classification

1. Benign/Indolent SFT (Grade I)

- Typical SFTs with low cellularity and minimal atypia.
- **Characteristics**:
    1. Spindle-shaped cells arranged in a “patternless” or storiform pattern.
    2. Presence of thick, ropy collagen bundles.
    3. Few mitotic figures (<4 per 10 high-power fields).
- Typically behave indolently with low recurrence rates.

2. Intermediate SFT (Grade II)

- Increased cellularity and moderate atypia without overt features of malignancy.
- **Characteristics**:
    1. Higher mitotic activity (4–9 per 10 high-power fields).
    2. Increased risk of recurrence and local invasion.

3. Malignant SFT (Grade III, Previously Hemangiopericytoma)

- High-grade tumors with aggressive behavior and metastatic potential.
- **Characteristics**:
    1. Marked cellular atypia and pleomorphism.
    2. High mitotic activity (≥10 per 10 high-power fields).
    3. Necrosis and vascular invasion often present.
    4. Rich vascular pattern resembling hemangiopericytoma in some cases.

Molecular Classification

- The hallmark molecular feature of all SFTs is the **NAB2-STAT6 gene fusion**, resulting from an intrachromosomal inversion on chromosome 12.
- **Implications**:
    1. Drives tumorigenesis through aberrant transcriptional activity.
    2. Causes nuclear overexpression of **STAT6**, which is a key diagnostic marker.

Anatomic Classification

1. Extrapleural SFTs

- Most common; arise in the soft tissues of the extremities, retroperitoneum, or head and neck.

2. Intrathoracic SFTs

- Originate from the pleura (classic site of SFTs).

3. CNS and Spinal SFTs

- Intradural, extramedullary lesions often located in the meninges or spinal canal.

WHO Grade and Behavior

- SFTs are classified within a single spectrum:
    1. **Grade I**: Benign with minimal recurrence risk after gross total resection.
    2. **Grade II**: Intermediate grade with higher recurrence risk but limited metastatic potential.
    3. **Grade III**: Malignant SFT (previously termed hemangiopericytoma), with high recurrence and metastasis rates.

Prognostic Factors

- **Histological Grade**:
    1. Higher-grade tumors (Grade II/III) are associated with poorer outcomes.
- **Mitotic Activity**:
    1. Higher mitotic rates correlate with increased recurrence and metastasis.
- **Resection Status**:
    1. Gross total resection (GTR) is associated with improved outcomes, especially in lower-grade tumors.
- **Adjuvant Therapy**:
    1. Radiotherapy is beneficial in cases of subtotal resection or high-grade lesions.

Clinical Applications

1. Diagnosis

- Confirmed with histopathology and STAT6 immunohistochemistry.
- Molecular testing can further validate NAB2-STAT6 fusion.

2. Treatment

- Surgical resection is the primary treatment.
- Radiotherapy is recommended for high-grade or incompletely resected cases.

3. Follow-Up

- Long-term surveillance is crucial, particularly for higher-grade tumors, due to potential for late recurrence.

The classification of SFTs underscores the importance of histological and molecular analysis in understanding tumor behavior and guiding management.

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