Diagnosis of Solitary Fibrous Tumors (SFTs)

The diagnosis of **solitary fibrous tumors (SFTs)** relies on a combination of clinical presentation, imaging studies, histopathology, and molecular analysis. Accurate diagnosis is critical due to their rarity and potential for variable behavior, ranging from benign to malignant.

Clinical Presentation

- **Symptoms**:
    1. Dependent on tumor location.
    2. **Spinal SFTs**:
        1. Spinal pain (most common).
        2. Neurological symptoms such as radiculopathy, motor weakness, or sensory deficits due to spinal cord or nerve root compression.
        3. Urinary dysfunction in advanced cases.
- **Onset**:
    1. Slow and progressive in most cases, with occasional acute presentations (e.g., hemorrhage or rapid growth).

Imaging Studies

- **Magnetic Resonance Imaging (MRI)**:
    1. Preferred modality for diagnosis and surgical planning.
    2. **T1-weighted images**: Iso- to hypointense compared to muscle.
    3. **T2-weighted images**: Iso- to hyperintense; may appear hypointense in highly fibrous tumors.
    4. **Contrast enhancement**: Homogeneous, intense enhancement after gadolinium administration.
    5. **Additional Features**:
        1. Displacement of adjacent structures (e.g., spinal cord compression).
        2. Dumbbell-shaped tumors in cases with foraminal extension.
- **Computed Tomography (CT)**:
    1. Useful for evaluating bony involvement and calcifications.
    2. Often used in conjunction with MRI for preoperative assessment.
- **Angiography**:
    1. May be utilized to evaluate tumor vascularity, particularly for highly vascular lesions.

Histopathology

- **Microscopic Features**:
    1. Spindle-shaped cells arranged in a “patternless” pattern.
    2. Alternating hypocellular and hypercellular areas.
    3. Thick collagen fibers.

4. High vascularity in hemangiopericytoma-like areas.

- **Grading**:
  1. Determined based on cellularity, mitotic activity, necrosis, and pleomorphism.
  2. Classified as Grade I (benign), Grade II (intermediate), or Grade III (malignant).

Immunohistochemistry

- **STAT6 Nuclear Staining**:
  1. Positive STAT6 staining is highly specific and diagnostic for SFTs.
- Additional Markers:
  1. Positive: CD34, Bcl-2, vimentin.
  2. Negative: S100 (to exclude schwannomas), EMA (to exclude meningiomas).

Molecular Testing

- **NAB2-STAT6 Gene Fusion**:
  1. A hallmark genetic alteration in SFTs.
  2. Confirmed using techniques such as next-generation sequencing (NGS) or fluorescence in situ hybridization (FISH).
- Molecular testing is especially useful in cases with atypical histology or immunohistochemical results.

Differential Diagnosis

- SFTs may be confused with other tumors due to overlapping features.
- Key differentials include:
  1. **Meningiomas**: EMA-positive, CD34-negative.
  2. **Schwannomas**: S100-positive, STAT6-negative.
  3. **Neurofibromas**: S100-positive with different histological patterns.
  4. **Sarcomas**: Higher grade, lack STAT6 staining.

Diagnostic Challenges

- Preoperative diagnosis is difficult due to the nonspecific clinical and imaging features.
- Diagnosis often requires histological and molecular confirmation post-resection.

Summary

The diagnosis of solitary fibrous tumors involves:

- **Imaging**: MRI with gadolinium is the gold standard.

- **Histopathology**: Patternless architecture and spindle cells.
- **Immunohistochemistry**: Positive STAT6 nuclear staining.
- **Molecular Testing**: NAB2-STAT6 gene fusion for confirmation.

Accurate and early diagnosis enables appropriate treatment planning, including surgical resection and, if necessary, adjuvant therapies.

Imaging

- **MRI (Preferred modality):**
  - Iso- to hypointense signal on T1-weighted images.
  - Variable signal on T2-weighted images (often hypointense due to fibrous content).
  - Strong, homogeneous enhancement with gadolinium.
  - May exhibit a “dural tail sign” similar to meningiomas.
- **CT Scan:**
  - Useful for detecting bone involvement or calcification.

Histopathology

- SFTs are composed of **spindle cells** arranged in a “patternless” pattern with alternating hypocellular and hypercellular areas.
- Immunohistochemical markers:
  - Positive: **STAT6**, CD34, Bcl-2, and vimentin.
  - Negative: S100 (helps differentiate from schwannomas or neurofibromas).