Spinal Solitary Fibrous Tumor

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A **spinal solitary fibrous tumor (SFT)** is a rare, usually benign, mesenchymal neoplasm that arises from the fibroblasts within the meninges or other connective tissue of the spine. Originally described as a tumor of the pleura, SFTs can occur in various locations, including the spinal canal, and are considered part of the spectrum of **extrapleural solitary fibrous tumors**.

Spinal hemangiopericytomas and spinal solitary fibrous tumors are now considered part of the same tumor spectrum under the unified classification of solitary fibrous tumors, with hemangiopericytoma representing a more aggressive, higher-grade variant. This understanding underscores the importance of molecular diagnostics and multidisciplinary management.

Key Features

Epidemiology

- Rare entity in the spinal region.
- Typically occurs in middle-aged adults but can present across all age groups.
- No significant gender predilection.

Pathogenesis

- SFTs arise from **mesenchymal fibroblastic cells** and are characterized by aberrant proliferation.
- Associated with the **NAB2-STAT6 gene fusion**, which is a diagnostic marker.

Clinical Presentation

- Symptoms often relate to **spinal cord compression** or **radiculopathy**, including:
  - Pain (local or radicular).
  - Neurological deficits such as weakness, sensory loss, or bowel and bladder dysfunction.
  - Paraparesis or quadriparesis in severe cases.
- Slowly progressive symptomatology is typical, but acute symptoms may occur with hemorrhage or sudden tumor growth.

Common Locations

- Intradural, extramedullary tumors predominate in the spinal canal.
- Thoracic spine is the most common site, followed by cervical and lumbar regions.

Diagnosis

Spinal Solitary Fibrous Tumor Diagnosis.

Differential diagnosis

spinal astrocytoma

hyperintense on T2 weighted images

ill-defined

patchy irregular contrast enhancement

eccentric location within the spinal cord

spinal ependymoma

hyperintense on T2 weighted images

hemorrhage is common

often associated with prominent cysts (tumoral and polar)

typically central with symmetric cord expansion

spinal ganglioglioma

hyperintense on T2 weighted images

mixed signal intensity on T1 weighted images

typically involve long segments of the spinal cord, often extending for greater than eight vertebral body segments

commonly eccentric in location

approximately half contain tumoral cysts

peritumoural edema is uncommon

patchy or no enhancement

calcification is common (low signal with blooming on GRE)

spinal hemangioblastoma

iso-hyperintense on T2 weighted images

focal flow voids on T2 weighted images

an associated tumor cyst or syrinx is common

hemosiderin capping may be present

spinal paraganglioma

hyperintense on T2 weighted images

usually located inferior to the conus

the characteristic “salt-and-pepper” appearance of neck and skull base paragangliomas may be seen

spinal intramedullary metastases

hyperintense on T2 weighted images

extensive peritumoural edema

Management

Surgical Resection

- **Gross total resection (GTR)** is the treatment of choice and is usually curative.
- Complete removal can be challenging if the tumor adheres to critical neurovascular structures.

Radiotherapy

- Considered for:

- Incomplete resections.
- Tumors with atypical features or malignancy.
- Recurrent disease.

Chemotherapy

- Rarely used, as SFTs are generally resistant to systemic therapy.
- Investigational for malignant forms.

Prognosis

- Generally favorable for benign SFTs with GTR.
- Recurrence is rare but possible, especially in incompletely resected or malignant cases.
- **Malignant transformation** occurs in a minority of cases, marked by increased mitotic activity, necrosis, or metastasis.

—- Prognosis of solitary fibrous tumors of the central nervous system remains unclear; consequently, careful and close monitoring of patients and long-term follow-up are mandatory. Radical surgical excision seems to be a significant and reliable prognosis factor, although pathological prognostic features must be defined. In other respects, the role of postoperative radiotherapy in atypical or incompletely resected solitary fibrous tumors of the central nervous system remains to be determined and, therefore, warrants larger series with longer follow-up periods [1].

Follow-Up

- Regular imaging (MRI) is recommended post-surgery to monitor for recurrence.
- Frequency varies based on resection status and histological findings.

Systematic review with original data

Apra et al. provide an up-to-date overview of their diagnosis, treatment, and prognosis. We included 10 primary STAT6-positive SFT from our retrospective cohort and 31 from a systematic review. Spinal pain was the most common symptom, in 69% of patients, and the only one in 34%, followed by spinal cord compression in 41%, radicular compression, including pain or deficit, in 36%, and urinary dysfunction specifically in 18%. Preoperative diagnosis was never obtained. Gross total resection was achieved in 71%, in the absence of spinal cord invasion or excessive bleeding. Histologically, they were 35% grade I, 25% grade II, and 40% grade III. Recurrence was observed in 43% after a mean 5.8 years (1 to 25). No significant risk factor was identified, but adjuvant radiotherapy improved the recurrence-free survival after subtotal resection. In conclusion, spinal SFT must be treated by neurosurgeons as part of a multidisciplinary team. Owing to their close relationship with the spinal cord, radiotherapy should be considered when gross total resection cannot be achieved, to lower the risk of recurrence [2]

Case reports

A 51-year-old male presenting with new-onset back pain and gait disturbances, who had undergone gross total resection (GTR) of an SFT within the thoracic spinal cord 19 years ago. Magnetic resonance imaging of the thoracic spine revealed recurrent tumors at the T7 level within the spinal cord. Subsequent resection achieved GTR. A comprehensive literature review was undertaken to assess the benefits of different resection extents (gross total removal (GTR) vs. subtotal removal (STR)), adjuvant radiation therapy, and the optimal duration of postoperative follow-up. Since 1960, 46 cases, including the present one, have reported recurrent spinal SFT/HPC following GTR and STR. Statistical analyses demonstrated that neither the type of resection nor adjuvant radiation therapy significantly impacted median recurrence-free survival in this cohort. Given their unpredictable behavior, meticulous lifelong follow-up following successful resection appears crucial for managing these tumors effectively [3].

A case of recurrent SFT of the lumbar spine with vertebral body involvement, presenting more than a decade after initial resection. It was initially misdiagnosed as a paraganglioma. To the best of our knowledge, there have been only 3 previous cases reporting SFT with vertebral body involvement [4]

Case Report from the HGUA

Patient Information

Age: 56 years **Gender**: Female **Transfer**: From another hospital **Religious Consideration**: Jehovah's Witness (signed refusal of blood products) **Allergies**: No RAMc **Toxic Habits**: Denies toxic habits

Medical History

- Hypothyroidism (on treatment with levothyroxine 75 mcg)
- Celiac disease
- Recurrent interstitial cystitis
- Bladder cysts
- Acute pyelonephritis (history)

Previous Surgeries

- Inguinal herniorrhaphy (right side)
- Appendectomy

Neurological Symptoms

- Pain in the right hypochondrium with progressive “numbness” in the abdomen and lower limbs.

- Difficulty walking with no reported motor deficits.

Neurological Examination

- **Consciousness**: Alert, oriented.
- **Cranial Nerves**: Intact.
- **Motor**: Mild weakness (5-/5) in proximal right leg (knee flexion-extension); otherwise normal strength.
- **Sensory**:
  - Paresthesias from T6 level downwards.
  - Thermal sensitivity: Normal.
  - Proprioception: Altered in the right lower limb.
  - Vibratory sense: 5/8 right medial malleolus, 6-7/8 left medial malleolus.
- **Reflexes**: Generalized, symmetric hyperreflexia. Bilateral Achilles clonus (exhaustible).
- **Pathological Reflexes**: Babinski positive on the right; flexor plantar response on the left.
- **Gait**: Cautious, slow with slight dragging of the right leg.

Imaging

- **MRI - Dorsal Spine**:
  - Intradural extramedullary lesion at D7 level (15 mm craniocaudal).
  - Bilobulated morphology with extension through the right radicular foramen.
  - Significant displacement and compression of the spinal cord to the left without signal changes.

Surgery

- **Procedure**: Laminoplasty and tumor excision.
- **Intraoperative Notes**:
  - Hemodynamic stability (TA: 110/70 mmHg; FC: 70 bpm; SatO2: 100%).
  - Preoxygenation and uneventful intubation.
  - Monitoring per SEDAR standards.
  - Blood loss: Within acceptable limits; no transfusion (Jehovah’s Witness).
  - Noradrenaline for low-dose hemodynamic support.
  - Total fluids administered: 1200 cc saline solution.
  - Postoperative Status: Extubated and transferred to UCCQ without complications.

Postoperative Course

- **Neurological**:
  - GCS: 15 points.
  - No new neurological deficits.
  - Mild residual paraparesis (4+/5) and hypesthesia in lower limbs.
  - Proprioceptive deficit in the right leg.
- **Rehabilitation**: Walking with a walker, undergoing physiotherapy.
- **General Status**:

- Afebrile.
- Dry, clean surgical wound.
- No signs of DVT.
- Tolerating oral intake and sitting position.

Outcome

- Favorable evolution during hospitalization.
- Discharged with follow-up in Neuro-Oncology outpatient clinic.

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1)
Metellus P, Bouvier C, Guyotat J, Fuentes S, Jouvet A, Vasiljevic A, Giorgi R, Dufour H, Grisoli F, Figarella-Branger D. Solitary fibrous tumors of the central nervous system: clinicopathological and therapeutic considerations of 18 cases. Neurosurgery. 2007 Apr;60(4):715-22; discussion 722. doi: 10.1227/01.NEU.0000255418.93678.AD. PMID: 17415209.
2)
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3)
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4)
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