CNS Lipoma

Definition and Classification

A central nervous system (CNS) lipoma is a rare, benign, slow-growing lesion composed of mature adipose tissue. Rather than being true neoplasms, they are generally considered developmental malformations (hamortomas) resulting from the abnormal differentiation of the meninx primitiva. They are categorized as CNS WHO grade 1.

Coding Identifiers:

- **ICD-O:** 8850/0

Subtypes and Variations

While histologically uniform, CNS lipomas are often categorized by their anatomical associations:

1. **Pericallosal Lipoma:** The most common type, frequently associated with dysgenesis of the corpus callosum.
2. **Spinal Lipoma:** Often associated with spinal dysraphism (e.g., spina bifida) or "tethered cord" syndrome.
3. **Quadrigeminal Plate Lipoma:** Located in the midbrain region.
4. **Cerebellopontine Angle (CPA) Lipoma:** Rare lesions that can involve cranial nerves.

Clinical Overview

- **Localization:** Most are midline structures. Common sites include the pericallosal region (50%), quadrigeminal/suprasellar cisterns, and the lumbosacral spine.
- **Presentation:** Many are asymptomatic and discovered incidentally. When symptomatic, they may cause seizures (pericallosal), gait disturbances, or myelopathy (spinal). CPA lipomas may cause hearing loss or vertigo.
- **Spread:** These lesions do not metastasize; they are benign and stay localized, though they can slowly expand or envelop adjacent neurovascular structures.
- **Epidemiology:** They account for less than 0.1% of all primary intracranial tumors. There is no strong gender predilection, and they are usually identified in children or young adults due to their congenital nature.

Etiology and Pathogenesis

- **Genetic Factors:** Most cases are sporadic. However, spinal lipomas are frequently part of complex congenital syndromes involving the neural tube.
- **Origins:** They arise from the persistent mesenchymal tissue that fails to resorb during the formation of the subarachnoid space.
- **Macroscopic Appearance:** They appear as soft, lobulated, yellow masses. They are often firmly adherent to or even encase surrounding nerves and blood vessels, making total surgical excision difficult and risky.

Diagnostics

Imaging

- **CT:** Pathognomonic appearance showing marked low density (fat density), typically measuring between -60 and -120 Hounsfield Units (HU).
- **MRI:** Characteristics follow subcutaneous fat:
    - **T1-weighted:** High signal intensity (bright).
    - **T2-weighted:** Intermediate to low signal intensity.
    - **Fat Suppression:** The signal completely "drops out" or disappears on fat-saturated sequences, confirming the diagnosis.

Pathology

- **Histology:** Composed of lobules of mature adipocytes (fat cells). They may contain varying amounts of collagenous connective tissue, small blood vessels, or occasionally calcification.
- **Immunophenotype:** Cells are positive for S100 and vimentin, consistent with mature fat cells.
- **Differential Diagnosis:** Must be distinguished from dermoid cysts (which often contain skin appendages and have different imaging signatures) and teratomas (which contain multiple germ layer tissues).

Management and Prognosis

Staging: As benign hamartomatous growths, there is no formal TNM staging system for CNS lipomas. They are classified by location and the presence of associated malformations.

Prognosis: The prognosis is generally excellent. Because these lesions are benign and grow very slowly (if at all), the clinical outlook depends primarily on the associated developmental brain or spinal defects rather than the lipoma itself.

- **Treatment:** Conservative management with serial imaging is preferred.

- **Surgery:** Surgical intervention is usually avoided unless there is significant mass effect or intractable symptoms, as the encasement of vital neurovascular structures makes complete removal dangerous.