Information Sheet on Superficial Siderosis (SS) for Neurologists

Introduction

Superficial siderosis (SS) is a rare neurodegenerative condition characterized by the deposition of hemosiderin, an iron-storage complex, in brain and spinal cord structures. While the progression of SS is typically slow, recent case studies have brought attention to an unconfirmed variant termed 'Acute Superficial Siderosis' where rapid progression occurs within a year. Understanding the disease's categorization, presentation, and management is pivotal in providing optimal patient care.

Categorization of Superficial Siderosis

1. **Infratentorial Superficial Siderosis (iSS) Type 1 (Classical)**:
    - **Localization**: Infratentorial regions, mainly involving the superior cerebellar vermis and at least one other infratentorial area.
    - **Radiological Significance**: The proximal VIII (vestibule-cochlear) cranial nerves are frequently involved but harder to visualize reliably.
    - **Etiology**: Likely due to chronic, low-volume, low-pressure extravasation of blood into the CSF over several decades.
2. **iSS Type 2 (Secondary)**:
    - **Localization**: Infratentorial, extensive involvement at the hemorrhage site and thin rim of hemosiderin in adjacent regions.
    - **Etiology**: Linked to a significant single acute intracranial hemorrhagic event.
3. **iSS Type 1 (Acute):**
    • Progression: Recently published case studies highlight a variant where disability progresses rapidly, 12 months from the triggering event to full disability. However, the exact cause of this swift progression remains undocumented and might be influenced by concurrent health conditions or comorbidities. Inclusion in this list is for informational purposes only.
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4. **Supratentorial Cortical Superficial Siderosis (cSS)**:
    - **Localization**: Supratentorial-only, with two variants:
        - **Focal**: Involvement of ≤3 sulci.
        - **Disseminated**: Involvement of >3 sulci.
    - **Etiology**: Associated with prior subarachnoid hemorrhage. In older individuals (>60 years), the primary cause is cerebral amyloid angiopathy (CAA).

CSF Analysis

- **Purpose**: To detect active or recent subarachnoid bleeding.
- **Key Markers**:
    - **Red Cell Count (RCC)**: Elevated RCC suggests recent bleeding. The highest levels are usually found in samples nearest to the hemorrhage site.
    - **Ferritin**: Increased in response to subarachnoid bleeding, potentially persisting for several months post-hemorrhage.

Neurological Management of iSS

1. **Addressing the Bleeding Source**:
    - **Goal**: Identify and treat any ongoing subarachnoid hemorrhage sources.
    - **Surgical Repair Considerations**: Suitable for patients who are good surgical candidates, have confirmed bleeding, show clinical progression, have a clear and treatable dural defect, and are likely to benefit more than the surgery's risks.
    - **Surgical Interventions**: Include repairing posterior fossa pseudomeningocele, spinal ventral dural defect, or cervical pseudomeningocele secondary to brachial plexus avulsion.
    - **Outcome**: Successful repair can lead to biochemical resolution in the CSF.
2. **Mitigating Neurotoxic Iron's Adverse Effects**:
    - **Strategy:** Use iron-chelating agents like deferiprone.
    - **Instruction:** Refer to the deferiprone dosing guideline specifically for SS patients.
    - **Note:** Patients undergoing chelation therapy should be monitored by hematology to ensure safe blood levels.

Concluding Notes

This information sheet serves as a starting point for neurologists unfamiliar with SS. While the definitive clinical benefits of certain treatments for SS remain undetermined, understanding its various forms and etiologies can guide diagnostics and interventions. Continuous patient monitoring is essential for advancing treatment strategies. It is crucial to approach each SS case with individualized care, considering the patient's overall health, disease progression, potential intervention benefits, and the clarity of the underlying cause.

Kharytaniuk, Natallia et al. “Classical infratentorial superficial siderosis of the central nervous system: pathophysiology, clinical features and management.” Practical neurology, practneurol-2021-003324. 11 Jul. 2022, doi:10.1136/practneurol-2021-003324