**Acquired Progressive Kinking of the Hair in the Setting of Parry-Romberg Syndrome**

**Jenna Beasley, M.D., Jason Sluzevich, M.D.**  
**Dermatology**  
**Mayo Clinic, Jacksonville, FL**

### Background

Parry-Romberg syndrome (PRS) results in unilateral facial atrophy of the skin and subcutaneous tissue with variable scarring alopecia and can clinically overlap with linear scleroderma.

Acquired progressive kinking of the hair refers to a number of conditions resulting in hair appearing curly, frizzy and lusterless. Localized involvement mimics a woolly hair nevus, but the majority of previously reported cases are postulated to reflect an early stage of androgenetic alopecia.

### Case History

67 year-old female with a remote history of prior sympathectomy who presented with a one year history of new-onset left hemi-facial atrophy associated with progressive curling of the hair involving the ipsilateral frontal-parietal scalp.

### Work up

- A punch biopsy of the involved face revealed mild dermal fibroplasia, focal lipoatrophy, and mild perivascular lymphocytic infiltrate but no well-developed dermal sclerosis.
- Magnetic resonance imaging confirmed atrophy of the subcutaneous adipose tissue with normal underlying fascia, muscle and bone.
- A punch biopsy of the scalp was non-inflammatory with preserved follicular units and normal telogen hair counts.
- Light and electron microscopy of the affected hair demonstrated mild cuticular weathering.

### Discussion

- An association between acquired progressive kinking of the hair and PRS has not previously been described in the indexed English literature. In this case, the hair kinking is temporally and anatomically related to the development of PRS, with the contralateral scalp hair showing no change. The absence of scarring alopecia and dermal sclerosis excludes linear scleroderma. The involvement of non-androgen dependent sites is inconsistent with conventional androgenetic alopecia. Ultrastructural hair examination is negative for a specific abnormality characteristic of an inherited keratin defect.
- The prior history of sympathectomy may be uniquely pathogenic, as there are established trophic effects of sympathetic innervation on hair maturation; additionally, sympathectomy has been documented to precede the development of PRS in both case reports and in animal models.

### References