Recurrent Schwannomatosis of the Hand: A Case Report and Relevant Literature Review

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BACKGROUND

1. Peripheral nerve sheath tumors (PNSTs) are neoplastic soft tissue masses generated from the abnormal proliferation of Schwann cells.
2. Often these tumors occur in isolation, “Schwannomas” or “neurilemmomas”, rarely as several “Schwannomatosis”
3. The purpose of this report is two-fold: (1) to review the relevant literature and describe a unique case of this rare condition, and (2) to emphasize salient clinical considerations in the diagnosis and treatment of Schwannomatosis

CASE

1. In this report we describe the case of a 52-year-old Caucasian male who presented with multiple recurrent soft tissue masses of the right hand.
2. On initial presentation he described pain across his right hand and index finger, which persisted despite numerous prior operations.
3. The index finger had a flexion contracture around the location of the proximal interphalangeal joint and there were multiple tender masses along the length of the finger and palm.
4. There were 3 prior surgical excisions, each followed by recurrence.

RESULTS

1. Segmental excision of the affected radial digital nerve was performed.
2. A pulp flap based on the contralateral neurovascular resulted in a sensate, pain free digit.
3. Tissue pathology confirmed the diagnosis of multiple neurilemmomas.

CONCLUSIONS

• This case illustrates an unusual presentation of Schwannomatosis.
• We emphasize the importance of:
  ➢ reviewing prior pathology and operative reports whenever performing repeat surgeries
  ➢ establishing a pre-operative differential diagnosis
  ➢ valuing preoperative imaging
• It was through careful consideration of surgical options and specific skin flap design, that this patient was able to retain a fully sensate, pain free, and mobile index finger post operatively after definitive hand surgery for Schwannomatosis.

Peripheral Nerve Sheath Tumor Differential Diagnosis

<table>
<thead>
<tr>
<th>Peripheral Tumor</th>
<th>Diagnostic signs</th>
<th>Other associated signs or symptoms</th>
<th>Inheritance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Solitary peripheral nerve sheath tumor</td>
<td>Intermittently painful, solitary lesion with non-aggressive features, + Tinel’s sign</td>
<td>If malignant: weight loss, night sweats</td>
<td>Sporadic</td>
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<tr>
<td>Schwannomatosis</td>
<td>Presence of two or more peripheral schwannomas with absence of clinical signs of Neurofibromatosis</td>
<td>Spares acoustic nerve</td>
<td>Familial (AD) or sporadic</td>
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<tr>
<td>Neurofibromatosis type 1 (NF 1)</td>
<td>Peripheral schwannoma, plexiform neurofibroma, Café-au-lait spots, Lisch nodules</td>
<td></td>
<td>Familial (AD)</td>
</tr>
<tr>
<td>Neurofibromatosis type 2 (NF 2)</td>
<td>Bilateral schwannomas affecting the acoustic nerve (cranial nerve 8), hearing loss</td>
<td>Cataracts, gliomas</td>
<td>Familial (AD)</td>
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