

Clinical Quiz

A painless finger mass

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Case

A 39-year-old gentleman presented with a painless finger mass that had increased in size over eight years. There was no history of trauma or injury. He denied having any numbness or tingling in his fingers. No similar masses were present elsewhere. Past medical and family histories were non-contributory. Physical examination revealed a non-tender soft mass on the extensor aspect of the right fifth middle phalanx which transilluminated (Figure 1A). Neurological examination including digital sensory and motor functions was normal. The radiograph showed a non-invasive soft tissue mass on the dorsal aspect of the fifth middle phalanx (Figure 1B). An aspiration attempt with an 18-gauge needle did not recover any fluid from the mass. The patient underwent a surgical resection by an orthopaedic hand surgeon. The extensor mechanism was noted to be attenuated with a large subtendinous encapsulated mass extending on the lateral sides of the digit. A 2 x 1 x 0.8 cm mass was carefully mobilized and completely excised. There was a large depression on the dorsal surface of the middle phalanx consistent with long-standing pressure on the bone with no invasive features.

Histological findings

On gross examination, the tumor was an encapsulated soft tissue mass with a light tan, homogeneous cut surface, without evidence of hemorrhage or cyst formation. Histologic examination of the excised specimen stained with hematoxylin-eosin revealed an encapsulated benign nerve sheath tumor. The mass was composed of spindle-shaped Schwann cells, elongated, wavy, and occasionally pointed nuclei and long eosinophilic processes without discernible cell borders predominately in an Antoni A pattern (Figure 2A). The lesion showed strong, dif-



Figure 1. A: Transillumination of the fifth finger mass. **B:** Radiograph of the fifth finger does not reveal any bone erosion.

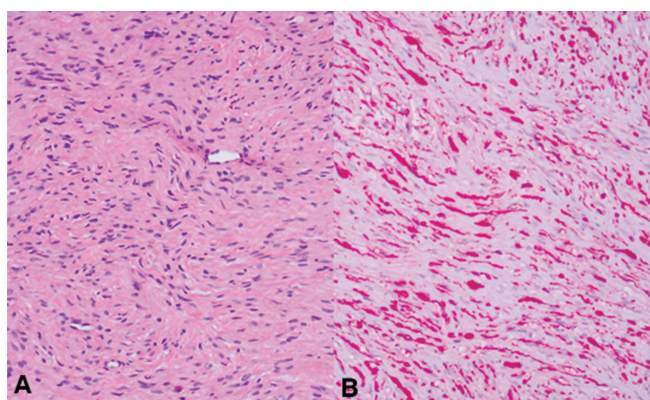


Figure 2. A: Histology reveals dense and highly fibrotic Antoni A (highly organized spindle cells with elongated and occasionally pointed nuclei and indistinctive cytoplasmic borders) tissue (hematoxylin and eosin, original magnification $\times 20$). **B:** Cells are positive for S100 (original magnification $\times 20$).

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fuse immunoreactivity for S100 protein (Figure 2B) and was negative for smooth muscle markers and CD34 consistent with the diagnosis of a digital nerve Schwannoma.

Commentary

Schwannoma (neurilemmoma) is the most common benign peripheral nerve tumor, yet it is rare accounting for less than 8 percent of all soft-tissue tumors¹. It is usually a firm nerve sheath tumor arising from the proliferation of differentiated Schwann cells from a single nerve funiculus^{1,2}. Schwannoma is usually a solitary, slow growing, and encapsulated tumor, and can occur anywhere in the body. Multiple schwannomas can be a part of neurofibromatosis (primarily in type 2) or as a syndrome called *schwannomatosis*². Bilateral vestibular schwannoma (VS) is the main manifestation of the neurofibromatosis type 2. Schwannomatosis is characterized by multiple spinal and peripheral nerve root schwannomas in the absence of VS. The most common reported locations of a sporadic schwannoma are the head and neck followed by the trunk and upper and lower extremities. It is less common in the wrist and hand region, occurring in approximately 1% of reported cases, and rarely occurs distal to the PIP joints¹. In the series reported by Rockwell et al.¹ only 4 of 18 wrist and hand schwannomas were located on the dorsum of the hand¹. Symptoms range from a painless mass to a painful mass causing paresthesia. Clinical diagnosis is difficult and it can be easily confused with ganglion cysts and other solid tumors including neurofibromas, lipomas, and giant-cell tumors^{3,4}. Transillumination may differentiate between cystic and solid lesions, although this procedure can be misleading in some cases⁴. As seen in Figure 1, not all transilluminating masses are ganglion cysts. Ganglion cyst is a benign mucin-filled cyst that remains connected to a joint capsule usually by a small calibre stalk. Diagnosis is usually made clinically. Diagnostic ultrasound is non-invasive and should be the initial diagnostic modality used to help differentiate solid from cystic masses¹. Presence of bone erosion on plain radiography raises the concern for invasive tumors. Needle aspiration with a large-bore needle can also help differentiate a ganglion cyst from a solid mass in the office setting. Magnetic resonance imaging (MRI) is a valuable diagnostic modality in evaluating soft tissue and bone structures of the hand¹. Although the diagnosis can be made by fine needle aspiration, an excisional biopsy is often necessary for definitive diagnosis and treatment. Nerve injury is the most significant complication of surgery.

Schwannomas are almost entirely composed of well differentiated Schwann cells. Usually areas of highly organized

spindle cells (Antoni A) alternate with less organized and hypocellular loose myxoid areas (Antoni B)¹. Well-formed palisading nuclei (known as Verocay bodies) that are important diagnostic features in schwannomas are usually absent in cellular schwannomas (uncommon schwannoma variant). Most schwannomas have a well-formed collagenous capsule. Thick and hyalinized vessel walls that may be seen in larger schwannomas are usually not present in cellular schwannomas. Large schwannomas which have undergone degenerative change (ancient changes) may show nuclear pleomorphism with areas of cyst formation, calcification, hyalinization, and hemorrhage. This should not be misinterpreted as an indicator of malignancy. Solitary neurofibromas may be confused with schwannomas. Schwannomas tend to grow on the side of nerve trunks and do not infiltrate into the nerve bundles, contrary to neurofibromas. Consequently, they can be completely enucleated. Histologically, neurofibromas do not demonstrate the alternating cellularity seen in schwannomas. Neurofibromas can also be immunopositive for S-100, although the positivity is usually less pronounced than in schwannomas^{1,2,5}. Schwannoma is usually surrounded by an EMA (epithelial membrane antigen) positive capsule. In contrast, the EMA is positive in only small number of neurofibromas mainly in perineurium-like structures⁵. In addition the CD34 staining is usually positive in neurofibroma^{1,2,5}. Interestingly, rare cases of hybrid peripheral nerve sheath tumor consist of neurofibroma and schwannoma have been reported². Malignant transformation is uncommon in schwannomas².

References

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Questions

1. Ganglion cysts are:

- A. The most frequent hand tumors
- B. Benign cystic lesions which can undergo a malignant transformation
- C. Usually painful
- D. Liquid-filled cystic lesions

Critique

The ganglion cyst is the most common hand tumor or tumor-like lesion. It is a benign cystic lesion and does not transform to malignancy. Ganglion cyst is usually painless, but sometimes it can be painful often when it is close to a nerve. It is a mucin-filled cyst that usually remains connected to a nearby joint capsule or a tendon sheath.

The correct answer is A.

2. What is the most common benign peripheral nerve tumor?

- A. Neurofibroma
- B. Schwannoma
- C. Lipoma
- D. Giant-cell tumor

Critique

Schwannoma is a rare benign peripheral nerve tumor which can occur anywhere in the body. Lipoma and giant-cell tumor are not peripheral nerve tumors. Neurofibroma is less frequent than schwannomas.

The correct answer is B.

3. Histologically, Schwannomas consist of:

- A. Schwann cells
- B. Antoni A
- C. Antoni B
- D. All of the above

Critique

Schwannomas are almost entirely composed of well differentiated Schwann cells. Usually areas of highly organized spindle cells (Antoni A) alternate with less organized and hypocellular loose myxoid areas (Antoni B). Well-formed palisading nuclei (known as Verocay bodies) are usually absent in cellular schwannomas.

The correct answer is D.

4. What is the most definitive way to diagnose a painless finger mass?

- A. Plain radiography
- B. MRI
- C. CT angiography
- D. Biopsy

Critique

Plain radiography can be helpful in detecting lytic bone lesions, but a normal radiograph does not exclude malignancy. MRI is a valuable modality in evaluating soft tissue and bone structures; however, an incisional or excisional biopsy is often necessary for a definitive diagnosis.

The correct answer is D.