Glomus Tumor

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Patient’s History

- J.C. is a 58-year-old male with a long history of left hand pain, sharp in nature, with occasional radiation to the left elbow and left shoulder. Pain is exacerbated by any activity of the left upper extremity.

- Patient has received a trial of non-steroidal anti-inflammatories, Neurontin, Elavil and Ultram, with transient relief.

- Physical exam - no skin changes, no temperature changes, no swelling and bilateral pulses.

- Neurological exam - no motor weakness, no sensory deficit, normal reflexes and mild hypersensitivity to light touch on the left hand.
Patient’s Work up

Patient underwent multiple diagnostic studies including:

- **Electromyogram (EMG)** of the left upper extremity - negative
- **Magnetic Resonance Imaging (MRI)** of the shoulder and cervical spine - unremarkable
- **Left hand X-ray** - *radiolucent lesion* of the distal tuft of the third digit.
Radiology Report

- 6 mm **radiolucent** lesion of the distal tuft of the 3rd digit
- **Thinning** of the adjacent cortex - risk for pathologic fracture
- **Sclerotic and well defined margins - benign** etiology
- **No other abnormalities of the bony structures**
Patient’s Work Up

• Pathology of soft tissue - aggregation of thick-walled blood vessels, most consistent with arterio-venous malformation, presence of **glomus bodies**.

• Based on clinical examination, X-ray findings and pathology, lesion was characterized as **glomus tumor** and subsequently excised.
What is Glomus Tumor?

• Rare benign vascular tumor, described by Wood in 1812

• Normal **glomerular unit:**
  - **neuromyoarterial apparatus** that serves as arteriovenous shunt
  - regulates **skin temperature**
  - found in the **reticular dermis** subunguially, on the finger tip pulp, on the base of the foot and the rest of body in descending order
Glomus Tumor

- **Extraosseus** lesions predominate
- **Subungual** site most common
- 75% of lesions occur in hand
- Other sites: palm, wrist, forearm, foot, stomach, mediastinum, eyelid, nasal fossa, colon and other

- **Bone involvement** - secondary manifestation due to invasion by a soft tissue glomus tumor

Clinical Presentation

- Occurs most frequently during the 4th or 5th decade of life
- Triad of sensitivity to cold, localized point tenderness and severe intermittent pain, described as burning or bursting
- Typically small lesions (< 1cm) in the soft tissue, not palpable or visible on physical exam
- Occasionally appears as localized pink or blue nodule beneath the finger nail
Diagnosis

• Standard approach to diagnosis is difficult because of small size and low prevalence

• Radiographic changes associated mainly with subungual tumors, but frequency does not exceed 50%-60%

• Some patients with painful symptoms may be considered to be psychiatric patients or malingerers

• 4-7 year period observed in routine practice that often precedes correct diagnosis
Radiographic Appearance

- **Soft tissue** glomus tumor produce shallow, well-marginated pressure erosions in the subjacent bone
- **Sclerotic margin** may be apparent due to slowly enlarging mass
- **Dorsal, medial or lateral surface in the tuft of a terminal phalanx** affected mostly

Daniel Wilner, Radiology of Bone Tumors and Allied Disorders, Sanders Company 1982: p769
Radiographic Appearance

- **Intraosseus** lesions appear as well-defined, osteolytic, cystlike lesion
- Underlying cancellous bone showing a sclerotic border
- Terminal phalanx predominate
Radiographic Appearance
Diff. Dx.

- Epidermal inclusion cyst
- Enchondroma
- Subungual carcinoma
- Keratoachanthoma
- Sarcoidosis
- Squamous Cell Carcinoma
Radiographic Appearance
Diff. Dx.

Epidermal Inclusion Cyst  Enchondroma  Subungual Carcinoma  Keratoacanthoma
Arteriography

- Initially method of choice
- Can be considered very invasive
- Presence of telangiectatic lake confirms the diagnosis

Courtesy of Drs. L. Ekelund and J. Gerlock, Inter. J. Radiol. 2:233, 1977
Magnetic Resonance Imaging (MRI)

- Limited spatial resolution and specificity of standard MRI techniques
- T1-weighted MRI - iso or slightly hyperintense to the dermal layers of the nail bed
- T2-weighted MRI - strongly hyperintense

Diff. Dx:
- mucoid cyst: high signal on T2; appearance on T1 similar to glomus tumor.
- angioma: same signal intensity as glomus tumor but more flat and superficial
A: Axial (a) and sagittal (b) views of subungual glomus tumor locations L=lateral, LF=lat fold, M=median, N=nail bed, NM=beneath the nail matrix, P=pulp, PF=proximal fold

B: (a) Axial T1-weighted MR image. (b) Axial T1-weighted MR image after administration of gadoterate meglumine. (c) Axial T2-weighted MR image

MR Angiography

- Contrast-induced T1-shortening effects in the vessels
- High contrast between the vessels of interest and the surrounding tissue.
- Glomus tumor appears as a small hypervascular area at the arterial phase

Ultrasound

- Reveals tumors as small as 3 mm
- Well-defined, hypoechoic, oval or round mass
- No differentiation between glomus and another small hypoechoic tumor
- Diff. Dx: epithelial, mycoid and synovial cyst, angiomas and hematomas

Fornage BD. Glomus Tumors in the Fingers: Diagnosis with US. Radiology 1988; 167:183-185
Histopathology

- Found at dermal and subdermal junction and has a fibrous capsule
- Various-sized vascular channels that are surrounded by masses of polyhedral glomus tumor cells (rounded uniform epithelioid cells with granular cytoplasm)
- Stain positive for smooth muscle marker actin

http://www.bonetumor.org/page138.html
Histopathology

Three forms of glomus tumor:

- **Vascular** form (glomangioma) – mostly vessels with little epithelioid component
- **Solid** form – sheets of glomus cells with few vessels
- **Myxoid** form – vascular channels and their cuff of cells are widely separated
Treatment

- Surgical excision
- Repair of nail bed after removal of subungual lesion
- Relief of pain intermediate after surgery
- Analgesics

Summary

- Rare, benign hamartoma arising from neuromyoarterial apparatus found in reticular dermis
- Triad of sensitivity to cold, localized point tenderness and severe intermittent pain
- Small size and low prevalence make standard approach to diagnosis difficult
- 50%-60% frequency of radiographic changes in subungual tumors
- Radiographic – shallow, well marginated pressure erosion in bone or osteolytic lesion, with sclerotic margin
- Arteriography – invasive choice – presence of telangiectatic lake
- MRI – rare use because of limited resolution – hyperintense on T2
- MRA – similar to arteriography – hypervascular area noted
- US – limited differentiation – well-defined hypoechoic area seen
- Diagnosis based on clinical picture, physical exam and radiographic changes
References

- Fornage BD. Glomus Tumors in the Fingers: Diagnosis with US. Radiology 1988; 167:183-185
- BIDMC patient records
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