Glomus Tumor Simulating Nerve Root Compression and Raynaud’s Phenomenon

Case Report

FRANK S. YELIN, M.D., AND EDMUND M. FOUNTAIN, M.D.

Division of Neurosurgery, Baylor University College of Medicine, and Methodist Hospital, Texas Medical Center, Houston, Texas

In 1812, William Wood of Edinburgh made the first good clinical description of lesions now thought to represent glomus tumors. He described these as “painful subcutaneous tubercles.” These painful nodules were probably known to ancient man as they are referred to in the writings of Hippocrates and Galen.

Campes (1760), Morgagni (1762), Bisset (1792), Dupuytren (1835), and Kolaczek (1878) have also referred to them. The latter interpreted this lesion as a variant of angiosarcoma. In 1920 Barre, a French neurologist, described a bluish subungual tumor associated with paroxysms of pain radiating proximally to the neck and sometimes coupled with a Horner’s syndrome and with changes in the vasomotor reflexes. Grosser in 1902 described the normal glomus body, but it was not until 1924 that Masson gave the first histological pattern of the tumor which arose from the neuromyoarterial glomus. The first reported cases in the American literature were in 1934 by Mason and Weil, and in 1935 by Raisman and Mayer.

Digital glomus tumors have often passed undiagnosed because of a lack of familiarity with them. We are presenting this paper to make the clinician more familiar with the historical, pathophysiological, and clinical aspects of this tumor.

The normal glomus is an arteriovenous shunt which is usually found in the corium of the fingertips. It may also be found about joints, the scapula, coccyx, stomach, and other less common sites such as the kidney, uterus, mediastinum, trachea, and bone.1,5,6 Histologically, it comprises an afferent artery and an efferent vein connected by a tortuous anastomotic vessel called the Sucquet-Hoyer canal. Lining the latter are endothelial cells, glomal cells, smooth muscle, collagenous material, and non-myelinated nerve fibers.4 The arterial blood flow is presumed to be controlled by the contractibility of the epithelial cells. The glomus is innervated by the autonomic nervous system. It is thought to be concerned with the mechanisms of regulation of local and general body temperature. The tumor is benign, although local infiltration may be present. When the glomus is subungual, the underlying bone may undergo pressure atrophy due to the pulsatile nature of this neuromyoarterial system (Fig. 1). The glomangioma is usually a solitary tumor and measures up to 8 mm in size.

Although these tumors are usually solitary, Kohout and Stout reviewed 731 cases and noted that multiple lesions in adults comprised 2.3%, while in children the incidence rose to 26.3%. The tumor is more common in males. Soule, et al., reported the incidence of glomus tumors to be 1.6% of 500 consecutive primary soft tissue tumors of the extremities.7 Glomus structures are absent in the newborn, but may be seen after the first year of life. The etiology of the tumor is unknown, and it has been stated that probably 50% appear after some trauma which may be minor.

Case Report

A 33-year-old woman was admitted to this hospital because for 13 years she had suffered from a sensitive spot on the ulnar side of the distal phalanx of the left index finger. This area was extremely painful to light touch, and the pain had become constant, with severe exacerbations on contact. For several months before admission the patient experienced pain radiating proximally from the sensitive finger to the left cervical region. She denied any motor weakness, paresthesias or other neurologic symptoms except the inability to use the painful and sensitive finger.

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Fig. 1. Photograph of the transillumination of our patient's normal and atrophic index fingers. Note the increased density in the region of the tumor, on the medial aspect of the atrophic digit.

Four years earlier the patient had had a "muscle sectioned and an artery cleaned off" in the left supraclavicular fossa without any therapeutic benefit. She subsequently tried paraffin baths, to no avail. Shortly thereafter she underwent the subtotal excision of three upper left ribs as well as the division of the left sympathetic trunk. A diagnosis of Raynaud's disease was made by the physician caring for her at the time. The pain persisted, and the remaining portion of a left cervical rib and scar tissue were excised with only temporary relief. The pain continued, and 6 months prior to admission, the patient received psychiatric evaluation and insulin shock therapy on two occasions. This treatment was discontinued as the patient became progressively more depressed and her pain worse.

Examination. The patient was a slender, cooperative, young woman. She had no carotid or supraclavicular bruises. Radial pulses were equal, and there was no evidence of a thoracic outlet syndrome. Healed scars in the left posterior chest and supraclavicular areas were noted. Reflexes were normal except for diminution of the left biceps reflex. Sensory modalities of touch, pain, vibration, position sense and stereognosis were normal except on the left index finger. The patient would not permit the examiner's finger to approach within 0.5 cm of the sensitive medial aspect of the distal phalanx. The patient held the sensitive finger in abduction; there was pulp atrophy over the terminal phalanx of this finger and a bluish discoloration of the skin on its medial portion. The remainder of the examination was normal including range of motion of the cervical spine.

Operation. A digital block of the involved finger was performed with total relief of the patient's symptoms. Transillumination of the involved finger revealed a nodule in the medial confines of the pulp space (Fig. 1). X-ray films of the phalanges were normal. The patient subsequently underwent surgical removal of a 0.5 cm bluish-purple mass from the involved phalanx. Six months postoperatively she remains symptom free.

Fig. 2. X-ray film showing osseous erosion of the distal phalanx of a finger, secondary to a glomus tumor. (Courtesy of Dr. R. H. Eppright, Houston, Texas)
Discussion

The clinical features of the glomus tumor should be well known. The tumor is small and has a bluish-purple or red hue, depending upon the degree of vascularity. It is exceedingly painful, and pressure may elicit paroxysms of pain with proximal radiation. The paroxysms may be fleeting or may persist for several minutes. Pain from a glomus tumor may be so severe that the patient may demand amputation of a limb or even contemplate suicide. Shugart, et al., compiled 74 cases and found that, as in our case, many patients refused to allow palpation of the lesion during examination.

When a tumor cannot be found for several months or years after the onset of pain, a diagnosis of a functional disorder often ensues. In a Mayo Clinic series, in only 43 of 74 cases was a tumor demonstrable on examination. Bony erosions may sometimes be seen in association with these tumors (Fig. 2). Vasomotor phenomena may exist, associated with the sympathetic nervous system. Cold usually accentuates and may initiate a painful paroxysm, although heat may act similarly. Transillumination may prove beneficial. Though not malignant, incomplete excision may lead to recurrence because of the local infiltrative nature of this tumor.

Summary

We have presented a case of a digital glomus tumor which was undiagnosed for 13 years and thus led to a variety of ineffective operations. Pain was completely relieved after diagnosis and complete excision of the tumor. Some of the historical, pathophysiological, and clinical features of glomus tumors have been noted.

References