

Deep Neck Hemangioma Masquerading as a Peripheral Nerve Sheath Tumor

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32-year-old man presented with a 1-year history of a nontender left level III neck lesion. He first noticed this "bump" in the mirror when turning his head to the right. It would occasionally swell with manipulation but had not become warm or tender and never exhibited discharge. An ultrasound performed by his primary care physician showed a 1.8-cm lesion deep to the platysma suggestive of a neurogenic lesion. On examination, a 2-cm nonpulsatile fullness of left level III was appreciated without an overlying punctum or dermal attachment. Magnetic resonance imaging (MRI) of the neck showed a well-circumscribed avidly enhancing oval nodule overlying the left sternocleidomastoid muscle but deep to the platysma, suggestive of a peripheral nerve sheath tumor (PNST; **Figure 1**).

After discussing observation vs surgical resection of a presumed neurogenic lesion of the superficial cervical plexus, he elected to proceed with surgical excision. In addition to the usual risks of excisional neck biopsy, including bleeding, infection, and scarring, he was counseled regarding possible weakness of the sternocleidomastoid muscle and overlying cutaneous numbness or paresthesia. An excisional biopsy was performed under general anesthesia. Intraoperatively, the lesion was multilobulated and well circumscribed. No contraction of surrounding musculature was noted with nerve stimulation.

Histologic sections showed a well-circumscribed proliferation of small- to medium-sized to scattered larger vessels. The vascular channels were irregular, dilated, and lined by a single layer of endothelial cells with absent to, at most, minimal atypia and no mitotic activity. The larger channels were separated by broad sclerotic septa. Lumina contained abundant blood and scattered fibrin thrombi. The features were consistent with a hemangioma (**Figure 2**).

This case report was determined to be exempt by the UCLA Institutional Review Board on July 30, 2021.



Figure 1. Postcontrast T1 magnetic resonance imaging of the neck shows an avidly enhancing well-circumscribed nodule (arrow) overlying the left sternocleidomastoid muscle, deep to the platysma.

Discussion

An initial suspicion of PNST can be, in part, justified by the low incidence of adult hemangioma as compared with PNSTs, which represent 12% of all benign soft tissue tumors. Given the typical pediatric presentation of hemangiomas, diagnosis in adulthood is less common. Of the few studies characterizing adult head and neck hemangiomas, one publication found that the majority of these lesions were localized

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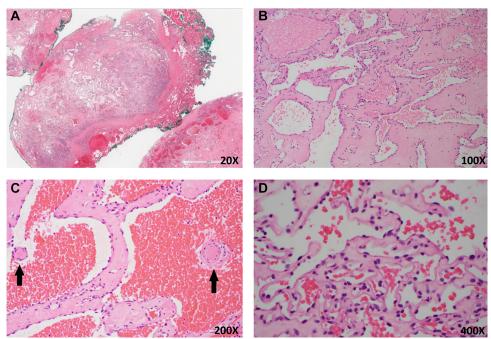


Figure 2. Histologic sections consistent with hemangioma: (A) well-circumscribed vascular proliferation, (B) irregular vascular channels, (C) scattered fibrin thrombi (arrows), and (D) channels lined by single layers of endothelial cells.

to the tongue, followed by the nasal cavity, parotid, and buccal region, and they were less likely to undergo spontaneous involution than pediatric hemangiomas.² As a result, most adult-onset hemangiomas are diagnosed after surgical excision.

Moreover, the radiographic findings of PNSTs and hemangiomas are similar. MRI scans of PNSTs and hemangiomas both exhibit T1 hypo- to isointensity and T2 hyperintensity. ^{1,3} In some instances, the type of PNST can be determined by characteristic features on MRI, as neurofibromas can exhibit a fascicular sign, while schwannomas can exhibit a target sign, but this is not always observed. ¹ Additionally, while neurofibromas and schwannomas can exhibit a "split-fat sign" seen on T1 due to a fine ring of fat around the lesion, malignant PNSTs often show notably heterogeneous enhancement on MRI. ¹ While hemangiomas have not been documented to exhibit a fascicular, target, or split-fat sign, they can show gadolinium enhancement, which can appear heterogeneous in areas of fat replacement, thereby mimicking some of the radiographic findings of PNSTs. ³

Upon histopathologic examination, PNSTs and hemangiomas can be readily differentiated. Hemangiomas are vascular neoplasms characterized by the proliferation of vessels of variable sizes, depending on subtype.³ In contrast, benign PNSTs such as schwannomas and neurofibromas are neoplasms with Schwann cell differentiation and demonstrate immunohistochemical staining for S100 protein.⁴

Given their low frequency in adulthood, hemangiomas are often diagnosed on pathologic examination. The difficulty in differentiating neck hemangiomas and PNSTs is not an isolated phenomenon, as challenges distinguishing internal auditory canal hemangiomas from vestibular schwannomas have

also been recorded.⁵ Although internal auditory canal hemangiomas may present with facial nerve dysfunction seemingly more often than vestibular schwannomas in early stages, misdiagnosis has occurred, only to be rectified upon pathologic evaluation.⁵ As they are similar radiographically to PNSTs, hemangiomas should be maintained on the clinician's differential when considering any indolent neck mass suspicious for a nerve sheath lesion. Patients should be counseled that definitive diagnosis may require pathologic analysis and that surgical excision of the tumor may be advised.

Author Contributions

Kristen A. Stefanescu, design and drafting of the manuscript, background literature review; critically revising the work for important intellectual content, approval of the final submission; agreement to be accountable for all aspects of the work; Chana R. Sachs, substantial contribution to the acquisition of data and drafting of the work, interpretation of pathology findings; critically revising the work for important intellectual content; approval of the final submission; agreement to be accountable for all aspects of the work; David Y. Lu, substantial contribution to the interpretation of pathology findings; critically revising the work for important intellectual content; approval of the final submission; agreement to be accountable for all aspects of the work; Ashley E. Kita, conception, design, and drafting of the work, acquisition of data, interpretation of data for the work; critically revising the work for important intellectual content; approval of the final submission; agreement to be accountable for all aspects of the work.

Disclosures

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