Unusual Tumor in an Interesting Site: Vascular Leiomyoma Originating from the Internal Jugular Vein: Case Report

A B S T R A C T

Angioleiomyoma is a benign tumor arising from the vascular smooth muscle (tunica media). Most of them are found in the uterus, skin and alimentary tract. It is rarely found in the head and neck area. Occurrences in the ear, the lip, the nasal cavity, the ethmoid sinus, the parotid gland and the larynx have been reported. We presented an unusual case of vascular leiomyoma originating from the left internal jugular vein in a 32 year-old man. To the best of our knowledge this is the first report of vascular leiomyoma originating from the internal jugular vein. All rare tumors like vascular leiomyoma should be kept in mind in the differential diagnosis of neck masses although physical examination findings are relevant with a simple benign mass.

Key Words: Angiomyoma; lymphangiomyoma; jugular veins

ÖZET


Anahtar Kelimeler: Anjiyomiyom; lenfanjiyomiyom; juguler ven

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Internal Jugular Ven Kökenli Vasküller Leiomyomi: Farklı Bir Bölgede Nadir Bir Tümör Olgu

Vascular leiomyoma (VL) is an uncommon benign tumor composed of smooth muscle cell and vascular endothelium. It originates from the tunica media of veins.\(^1\)\(^-\)\(^3\) They are also known as angiomyoma, angioleiomyoma or dermal angioma. It can occur anywhere in the body where smooth muscle is found.\(^2\) According to Enzinger and Weiss, 95% of 7748 leiomyomas occurred in the female genitalia, 3% in the skin, 1.5% in the gastrointestinal tract and the remainder in various other sites.\(^4\) Those in the head and neck area are rare and occur less than 1%.\(^5\) Occurrences in the ear, the lip, the nasal cavity, the ethmoid sinus, the parotid gland and the larynx have been reported.\(^6\)

In 1973, Morimoto classified angioleiomyomas into three histological types: solid, cavernous, and venous types. The solid type is more common
in females, and the venous type is more common in males. The cavernous type is the least common type of the three.\textsuperscript{2}

Clinically, VL presents as a solitary, small, painful cutaneous mass of the solid type.\textsuperscript{1} It causes pain in 60\% of patients.\textsuperscript{2} This tumor is rarely diagnosed before surgery. The usual treatment is excision of the mass and ligation of the feeding vessel.\textsuperscript{2} We present a case of a vascular leiomyoma which was adherent to the upper third of the internal jugular vein.

\section*{CASE REPORT}

A 32 year-old man presented with a 1 year history of a slowly enlarging nontender left neck swelling. Physical examination showed a 3x2.5 cm. mass located along the anterior edge of the left sternoclaviodomastoid muscle near the angle of mandible. It was smooth, firm, mobile and nontender. There were no abnormal findings in the head and neck examination. Computed tomography scan with contrast showed an enhanced mass located at the bifurcation of the left carotid artery (Figure 1). Due to its perceived vascular origin, MR angiography was performed. It showed that the mass was not vascular (Figure 2 ). So it was thought as a simple benign mass and excisional biopsy was planned under general anesthesia. During surgery, the solitary mass was found to be adherent to the adventitia of the left internal jugular vein at the level of bifurcation of the carotid artery. The mass and its capsule were delicately dissected from the internal jugular vein, but the jugular vein needed to be repaired because of an injury. There was no postoperative complication. No recurrence is observed.

\section*{PATHOLOGY}

Grossly the specimen showed an ovoid, firm mass measuring 1.5x1x0.5 cm. The cut surface was solid and pale tan. Histologically, it consisted of a well demarcated nodule of smooth muscle tissue, punctuated with thick walled vessels with partially patent lumina. The inner layers of vessels' smooth muscle were arranged in an orderly circumferential fashion, the outer layers were arranged perpendicular to the inner layers. Areas of myxoid change were seen. The vessels within these tumors lacked internal and external elastic lamina. The tumor was made up predominantly venous vessels, consistent with a venous type of angioleiomyoma. (Figures 3 and 4).

Immunohistochemistry using a peroxidase-antiperoxidase method (PAP) for both alpha smooth muscle actin and CD34 tested positive, confirming the diagnosis of venous type angioleiomyoma. (Figures 5 and 6)

\section*{DISCUSSION}

Leiomyomas are rare, benign, soft tissue tumors arising from smooth muscle. Smooth muscle tumors arising from the muscle layer of blood vessels are known as vascular leiomyoma or angioleiomyo-
It most commonly arises from the smooth muscle of the female genital tract. The next most common site is smooth muscle of the gastrointestinal system. Of all leiomyomas in the human body, less than 1% occur in the head and neck area. Vascular leiomyomas are even less common.

VL can develop in the oral or nasal mucosa, larynx, deep spaces such as parotid space, submandibular space, carotid sheath, facial bones or retropharyngeal space. The auricle and oral and sinonasal cavities are the most commonly reported sites of occurrence in the head and neck.

Most commonly, patients with a leiomyoma present with a slowly enlarging mass that sometimes may be tender. In our case, there was a 1 year history of a slowly enlarging, nontender mass.

The histogenesis is unclear. Duhig and Ayer proposed that proliferation of smooth muscle wit-
hin a hemangioma produces a vascular leiomyoma and further proliferation produces a simple leiomyoma. This may explain why some of these tumors may be hypervascular, whereas the more common leiomyoma is more solid and less vascular. It is possible that the lesion may represent a hamartoma rather than a true neoplasm.

On histological examination, smooth muscle bundles and vascular channels surrounded by a thin capsule are seen. Numerous vessels of various sizes, having muscular walls of varying thickness were encompassed by interlacing bundles of smooth muscle. Muscle cells were disposed tangentially from the periphery of the blood vessels to merge with and form a part of the intervascular muscle fascicles. The stroma contained varying amounts of collagen, indicating myxomatous degeneration. Fine needle aspiration is not sufficient to diagnose angioleiomyoma. So, the diagnosis of a leiomyoma is often difficult to make before a histological examination.

In the case we present, the histological diagnosis was done after excisional biopsy.

Morimoto classified these tumors into three histologic subtypes: solid or capillary, cavernous, and venous. The most common type found in the head and neck area is the venous type. The solid type has smooth muscle bundles that intertwine and surrounds the vascular channels. The cavernous type has dilated vascular channels with less smooth muscle. The venous type has vascular channels with thick muscular walls that are easily discerned from smooth muscle bundles.

The presence of pain is related to the histological type of the tumor. The solid type is usually painful, and the venous or cavernous type is painless. If a vascular leiomyoma is deeply seated, it can grow large because of the absence of symptoms. But most vascular leiomyomas are small in size due to their superficial location and are slow growing in nature. Hasegawa et al. suggested that the pain in these tumors may be mediated by the nerve fibers located within the tumor parenchyma. Small nerve fibers were identified within the stroma of 69% of painful tumors.

Malignant variants of this neoplasm have been reported in rare cases. The distinction between benign and malignant smooth muscle tumors in extraterine sites can be difficult. Size, infiltration, bone erosion in significant sites and mitotic index are valuable diagnostic aids. The most useful histologic indicator of a benign lesion is the absence of mitoses. Several stains such as desmin, vimentin, Masson’s trichrome, actin and myosin have been used to identify vascular leiomyomas. However, this spectrum of stains typically is not required for the diagnosis.

Fine needle aspiration cytological examination and imaging studies such as sonography, computed tomography (CT), or magnetic resonance imaging (MRI) have no characteristic findings for preoperative diagnosis. Surgical excision with histological analysis is the only way to make a diagnosis.

The histopathologic differential diagnosis for these lesions includes hemangiomias, nasal angiofibromas, fibromyoma, leiomyoblastoma, angiomylipoma, and vascular leiomyosarcoma. Only one case of vascular leiomyoma located at the carotid bifurcation has been reported by Reiner et al. Our case is the first report of vascular leiomyoma originating from the internal jugular vein.

Although very rarely seen, vascular leiomyoma must be kept in mind in the differential diagnosis of head and neck masses. Simple excision biopsy is often curative. Recurrence rates are reported to be very low. In the review by Hachisuga et al., only 2 of 562 angioleiomyomas recurred. In our case, there was no recurrence within two years.
REFERENCES