Mixed Lymphangioma and Cavernous Hemangioma Within the Ulnar Nerve: A Case Report

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Abstract

Background: Lymphangiomas are benign lymphatic vessel hamartomas typically found in the skin or subcutaneous tissue of the head and neck. Although mostly seen in a congenital context, acquired forms have been reported. By contrast, cavernous hemangiomas are benign hamartomas of endothelial origin. They can arise anywhere in the body, but are typically described as arising from the central nervous system. **Methods:** We report the case of a young patient who developed a mixed lymphangioma and cavernous hemangioma within the ulnar nerve. The tumor was removed during an intrafascicular dissection with preservation of all major fascicles. **Results:** Postoperatively, the patient retained intrinsic motor function and full sensation returned. This is the first published case of this type of mixed tumor occurring in a peripheral nerve. **Conclusions:** It could not be ascertained whether trauma was the mechanism by which a lymphangioma developed within the peripheral nerve, as has been proposed. Although this is plausible, alternative mechanisms should be considered.

Keywords: lymphangioma, mixed lymphangioma, cavernous hemangioma, hemangioma, ulnar nerve

Introduction

Lymphangiomas, or benign lymphatic vessel hamartomas, are typically seen in the skin or subcutaneous tissue of the head and neck region.³ Although most often described in a congenital context, acquired forms, or lymphangiectases, are known to occur, usually as the result of scarring from surgery or radiation therapy. Acquired lymphangiomas are rare in the peripheral nervous tissue, with few previous cases reported in the literature.^{5,7,12,13} It is speculated that lymphangiomas that arise in peripheral nerves are traumatic in origin.¹²

Cavernous hemangiomas constitute a rare form of hemangioma. Although they are most frequently described as arising in the central nervous system, they have also been reported in ectopic locations.^{2,8,9} No previous reports of cavernous hemangiomas arising in the distal peripheral nerves have been found. We report the case of a young patient who developed a mixed lymphangioma and cavernous hemangioma in the ulnar nerve. This is the first published case of this type of mixed tumor occurring in a peripheral nerve.

Case Report

The patient is an 18-year-old, right-hand-dominant female who initially presented to her primary care provider with a 1-year history of a painful mass in the volar ulnar aspect of her left wrist. Over the preceding 6 months, the mass had

become more painful; a palpable lesion had developed that grew in size and caused increasing discomfort. The patient additionally complained of sporadic paresthesias in the fourth and fifth digits of the left hand. She was seen by a neurologist, and a magnetic resonance imaging (MRI) was obtained which showed a lesion thought to be consistent with a schwannoma. The patient was referred for surgical evaluation.

A complete medical history review was unremarkable. Physical examination revealed normal motor and sensory exams in all extremities. No epitrochlear or axillary lymphadenopathy was noted. A 1-cm mass was present on the volar aspect of the ulnar side of the left wrist. Distal tingling to percussion (Tinel sign) was noted over the fourth and fifth digits over the area of the mass. The patient also had a tattoo directly over the surface of the mass. Surgical biopsy and removal of the mass, with the possibility of multistage removal if the biopsy revealed an aggressive tumor, were planned.

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Surgical exploration revealed a 1.0 cm × 0.7 cm mass within the ulnar nerve just proximal to Guyon canal. Significant bleeding and fluid egress from the lesion, inconsistent with a schwannoma, were noted. A biopsy was obtained revealing large, irregular vascular spaces lined by flattened, bland epithelial cells as well as lymphatic channels consistent with an intraneural mixed lymphangioma and cavernous hemangioma. There was no evidence of malignancy. Complete removal of the mass was recommended. Due to the tumor's intrafascicular involvement within the ulnar nerve, a microsurgical consultation was also obtained prior to removal of the mass. The tumor was then removed during an intrafascicular dissection with preservation of all major fascicles. Postoperatively, the patient retained intrinsic motor function and full sensation. At 3-month follow-up, she reported that the sporadic paresthesias in the fourth and fifth digits of the left hand had resolved. One year postoperatively, the patient was doing well with no signs of reoccurrence.

Discussion

The most common types of lymphangiomas occur congenitally, often seen clinically as lymphangioma circumscriptum, cavernous lymphangiomas, and cystic hygromas. In addition, these forms can be viewed as part of common congenital syndromes with karyotype abnormalities such as Down syndrome, Turner syndrome, Noonan syndrome, and trisomies 13 and 18.³ Lymphangiectases may result from inflammation or the fibrosis of lymphatic channels leading to an accumulation of lymphatic tissue, such as are noted following surgical procedures or radiation therapy.

The etiology of acquired peripheral nerve lymphangiomas is difficult to ascertain, and there is relatively little published literature on the subject. Sun et al¹² have speculated that because of the exorbitant secretion of lymph from damaged endothelial cells or decreased drainage from scarred lymphatic channels, trauma plays a major role in the development of peripheral nerve lymphangiomas. In this case, the patient did not have a history of trauma to the left wrist, though she did have a tattoo directly over the surface of the mass.

Conclusion

Although hemangiomas have been noted to arise in peripheral nervous tissue, most reports detail their appearance in nerve roots, retrobulbar locations, and the cranial nerves.^{10,11} There have been few reports of hemangiomas arising in the distal peripheral nerves, including the radial nerve and the digital nerves.^{4,9} Cavernous hemangiomas, which constitute a subtype of hemangioma, arise in the central nervous system. Structurally, they are dilated blood vessels consisting of a single layer of endothelium. Although extra-axial cavernous hemangiomas have been

reported,^{6,9} none have been reported as occurring in nerves other than cranial ones.^{1,4}

To our knowledge, this is the first published report of a cavernous hemangioma arising within the ulnar nerve, as well as the first report of a mixed lymphangioma and cavernous hemangioma arising in nervous tissue. In this case, it could not be ascertained whether trauma was the mechanism by which a lymphangioma developed within the peripheral nerve, as has been proposed. Although this is plausible, alternative mechanisms should be considered.

Ethical Approval

This study was approved by our institutional review board.

Statement of Human and Animal Rights

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008. Informed consent for inclusion in the study was obtained from all patients.

Statement of Informed Consent

Even though there were no identifying details about the patient in this study, informed consent was obtained for inclusion in the study.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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