Surgical Oncology & Clinical Practice Journal

Editorial
Published: 21 Dec, 2016

Nerve Hamartomas - Fact or Fiction?

Jenna-Lynn Senger[1] and Rani Kanthan[2]**

[1]Division of Plastic Surgery, University of Alberta, Canada

[2]Department of Pathology and Laboratory Medicine, University of Saskatchewan, Canada

Editorial

Hamartoma, derived from the Greek hamartia meaning "failure", is a benign malformation composed of local native tissues with disorganized growth. Hamartomas are the most common benign tumors of the lung and the pediatric heart (cardiac rhabdomyomas), though they can be found throughout the body in most anatomical structures. Though typically benign 'incidentalomas', they can cause adjacent compression of critical vessels and nerves, resulting in a myriad of clinical presentations. Within the peripheral nervous system, two types of nerve hamartomas have been described: a) fibrolipomatous hamartomas, and b) neuromuscular hamartomas.

a) Fibrolipomatous hamartomas (FLH) (aka lipomatous hamartoma, perineural lipoma, intraneural lipoma) are rare fibrofatty malformations of the peripheral nerve that most often arise in the first three decades of life, with more than 80% arising in the median nerve [1]. FLH have also been reported in digital, ulnar, radial, sciatic, peroneal, cranial nerves and brachial plexus [2,3]. These hamartomas form due to the overgrowth of the fibroadipose tissue surrounding and within nerve bundles, causing nerve enlargement [1]. Symptoms are therefore those of a compressive neuropathy, with carpal tunnel syndrome being the most common in afflictions of the median nerve. Bilateral cases of median nerve FLH are extremely rare [3]. Though most cases are isolated, up to one-third will present with macrodactyly; fibrolipomatous hamartoma in the setting of macrodactyly is termed 'macrodystrophia lipomatosa' [1]. Nerve conduction tests and electromyograms [EMGs] demonstrate sensory and motor disturbances; both ultrasound and MRI imaging show pathognomonic coaxial cable appearance [2]. Accurate histopathological diagnosis is challenging, as FLH has mature adipose and connective tissues that is intimately admixed with the nerve fibers by surrounding and infiltrating the nerve [1]. The differential diagnosis is thus large, and includes lipomatous neurofibroma, lipofibromatosis, lipomatosis, intraneural perineurioma [1].

b) Neuromuscular hamartomas (NMH) (aka neuromuscular choristomas, nerve rhabdomyoma, benign triton tumors) are rare benign intraneural tumors characterized by the intimate admixture of mature skeletal elements admixed with mature neural elements and no cellular atypia or atypical mitoses. Such hamartomas are hypothesized to arise from one of four mechanisms: i) muscle spindle hamartomatous growth, ii) skeletal muscle entrapment within the nerve during embryogenesis, iii) neuroectodermal differentiation into skeletal muscle, or iv) epigenetic alteration of motor end plates [4]. Similar to FLH, they most often present as a solitary lesion in childhood and have been commonly reported to occur as an enlarged fusiform mass in large nerves such as the sciatic and median nerves as well as the brachial plexus [5,6]. Rare accounts of these NMHs arising in obscure locations such as the head and neck have also been reported [4]. When involving the sciatic nerve, patients present with progressive limb deformity and neurologic deficits [5]. MRI is often nonspecific, showing a lesion of low to intermediate signal-intensity T1WI that may demonstrate fibromatosis. Though categorized as a 'hamartoma' and thus a benign entity, NMH are classified based on their location as it dictates the tumor aggressiveness: peripheral lesions are not aggressive, as one would expect with a hamartoma, yet central lesions are biologically aggressive with an infiltrative destructive growth pattern [4]. Given the natural history of these tumors, one has to question whether 'nerve hamartoma' is a correct classification for these aggressive lesions. Malignant triton/NMH tumors are characterized by benign epithelial elements admixed with rhabdomyosarcomatous mesenchymal differentiation on a background of Schwann cells [7]. Perhaps the 'aggressive' NMHs represent an intermediate spectrum between the benign and malignant triton tumors; though by definition if the potential for malignant degeneration exists, such lesions should not be termed 'hamartoma'.

Nerve hamartomas are rare lesions that are often clinically silent and, when symptomatic, are easily misdiagnosed. As such, there remain no consensus-based guidelines for management, with

OPEN ACCESS

***Correspondence:**
Jenna-Lynn Senger, Division of Plastic Surgery, University of Alberta, 5005C Katz Group Centre, Edmonton, Alberta, T6G-2E1, Canada,
E-mail: jennalynn@ualberta.ca
Rani Kanthan, Department of Pathology & Laboratory Medicine, Royal University Hospital, 103 Hospital Drive, University of Saskatchewan, Saskatoon, SK S7N 0W8, Canada,
E-mail: rani.kanthan@saskatoonhealthregion.ca

Received Date: *06 Dec 2016*
Accepted Date: *19 Dec 2016*
Published Date: *21 Dec 2016*

Citation:
Senger J-L, Kanthan R. Nerve Hamartomas - Fact or Fiction?. Surg Oncol Clin Pract J. 2016; 1(1): 1004.

available literature being retrospective, anecdotal and predominantly case based reports. Treatment options of these rare nerve lesions range from conservative observation to invasive complete nerve resection. Most peripheral nerve surgeons advocate for nerve decompression in symptomatic patients with nerve biopsy to rule out malignancy. While most patients respond well to simple decompression, a select few will have recurrences that may require aggressive resection with an interpositional nerve graft [2]. If the motor branch is nonfunctional, or a long nerve graft is required, reconstruction using a distal nerve transfer should be considered. The use of aggressive resection as a first-line treatment, including epineurotomy and intraneural dissection, remains controversial in the published literature, with some authors reporting excellent results, though associated with the high risk of the significant morbidity of neuronal loss of sensory and/or motor function. As such, many advocate for these drastic resection measures to be reserved for repeated recurrences or when the nerve function deteriorates to the point where the possible morbidity of surgery is less than the morbidity of no treatment [2]. In the case of "benign" triton [NMH] tumors, aggressive excision is offered as the first line of management for the aggressive central type [4].

In conclusion, nerve hamartomas though extremely rare, do exist and are a real fact and not fiction. The natural history of these lesions remains largely unknown and is not fully characterized. These lesions are ascribed to be responsible as some of the rare causes of compression neuropathy with the potential to irreversibly influence motor and/or sensory function. Lipofibromatous hamartomas are benign tumors composed of fibrofatty tissue overgrowth and are therefore a "factual" hamartoma; however, accuracy of calling benign triton tumors a type of 'hamartoma' given the propensity of a subset for destructive growth is questionable. Despite their similar clinical pictures, differences in the natural history of these two tumor- types suggest the extent of surgical management should differ. Additionally, steps for the future may include revision in nomenclature with subsequent development of individualized treatment guidelines to ensure proper weighing of the risk of surgical morbidity with risk of tumor progression for best outcomes.

References

1. Plaza JA, Kazakov DV, Casas G, Requena L, Sanmartin O, Kacerovska D, et al. Fibrolipomatous hamartoma of the nerve: a clinicopathological report of 13 cases. J Am Acad Dermatol. 2014; 70: 736-742.
2. Agarwal S, Haase SC. Lipofibromatous hamartoma of the median nerve. J Hand Surg. 2013; 38: 392-397.
3. Senger JL, Classen D, Bruce G, Kanthan R. Fibrolipomatous hamartoma of the median nerve: a cause of acute bilateral carpal tunnel syndrome in a three-year-old child: A case report and comprehensive literature review. Plast Surg(Oakv). 2014; 22: 201-206.
4. Bae DH, Kim CH, Cheong JH, Kim JM. Adulthood benign triton tumor developed in the orbit. J Korean Neurosurg Soc. 2014; 56: 146-148.
5. Lam S, Grandhi R, Wong R, Hamilton R, Greene S. Neuromuscular hamartoma of the sciatic nerve: case report and review of the literature. Surg Neurol Int. 2013; 4: 8.
6. Uysal A, Sungur N, Kocer U, Cöloğlu H, Oruç M, Yalta T. Neuromuscular hamartoma of the occipital nerve: clinical report. J Craniofacial Surg. 2005; 16: 740-742.
7. Senger JL, Kanthan SC, Kanthan R. Sinonasal malignant triton tumor. A case report with historical & surgical review. Head and Neck Cancer Res. 2016; 1: 2.