



Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Nerve Sheath Myxoma: A rare tumor, a case report and literature review

Haitham Khashaba^{a,*}, Eman Hafez^b, Hisham Burezq^c^a University Hospital of North Durham, Durham, UK^b Royal Victoria Infirmary, Newcastle Upon Tyne, UK^c Al Babbain Burns, Plastic and Reconstructive Surgery Center, Kuwait

ARTICLE INFO

Article history:

Received 13 May 2020

Received in revised form 7 July 2020

Accepted 8 July 2020

Available online 15 July 2020

Keywords:

Hand swellings in children

Nerve sheath myxoma

Nerve sheath tumors in children

Neurothekeoma

ABSTRACT

INTRODUCTION: Nerve Sheath Myxoma is a rare benign nerve sheath tumor, commonly seen in young adults. Patients usually present with a small nodule commonly affecting upper and lower limbs. Nerve Sheath Myxoma has not been reported in children below the age of 1 year.

CASE PRESENTATION AND METHODS: We present the first reported case of Nerve Sheath Myxoma (NSM) in the hand in a 4-month old child in line with the SCARE 2018 criteria.

DISCUSSION: NSM is difficult to diagnose using standard imaging technique; Magnetic Resonance Imaging (MRI) and Ultrasonography (USS) cannot differentiate it from vascular anomalies.

CONCLUSION: Soft tissue tumors in the hand of pediatric population are difficult to diagnose. Formal excision of the tumor by a specialized pediatric hand surgeon and histopathological analysis were the only avenue to correctly diagnose the NSM.

© 2020 The Author(s). Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

Nerve Sheath Myxoma (NSM) is a rare benign tumor of the perineural “Schwann” cells of peripheral nerves [1]; commonly affects the limbs of young adults with a peak incidence in the thirties [2]. It was first described by Harkin and Reed in 1969 [3]. Previously, the term Neurothekeoma that was first introduced by Gallager and Helwig in 1980 [4], was considered as a variant of NSM [2]. Recently Sheth and colleagues clearly differentiated between both entities; Nerve Sheath Myxoma and Neurothekeoma, regarding genetic expression and cell origin [5].

2. Methods

In line with the SCARE criteria [18], we report a case of Nerve Sheath Myxoma (NSM) in a 4-month old female infant. We discuss the presentation, clinical picture, radiological studies, and histopathology leading to the final diagnosis



Fig. 1. Preoperative picture showing 3 × 3 cm swelling in the right thenar eminence.

3. Case report

A 4-month old girl was presented to our clinic with a lemon size swelling in the right hand of 1-month duration, slowly growing, with no history of trauma reported by parents.

On examination, the infant had a 3 × 3 cm well circumscribed, non-tender subcutaneous swelling in the right thenar eminence (Fig. 1). The swelling was soft, smoothly lobulated, non-pulsating and partially attached to the overlying skin but was not attached to

* Corresponding author.

E-mail addresses: Haitham.khashaba@nhs.net (H. Khashaba), Eman.hafez@nhs.net (E. Hafez), burezq@gmail.com (H. Burezq).

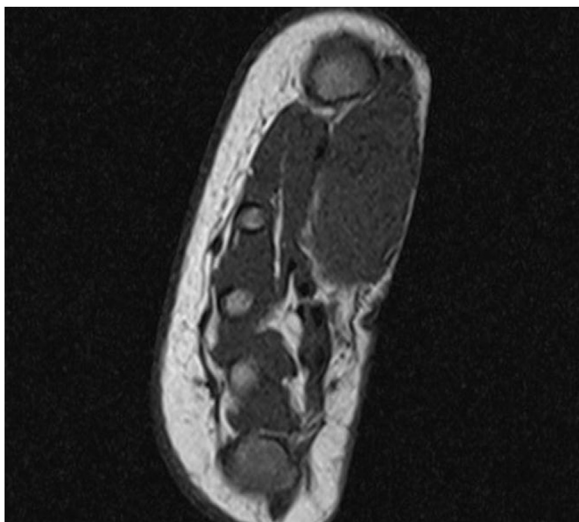


Fig. 2. Axial T1 weighted image of the right hand demonstrating a well-defined subcutaneous mass isointense to the muscle in the thenar eminence.



Fig. 3. Axial T1 weighted - short tau inversion recovery (STIR) image of the right hand - demonstrating a well-defined subcutaneous hyperintense mass in the thenar eminence.

the underlying structures. There was no affection of hand or thumb movement.

Duplex Ultrasonography showed a subcutaneous circumscribed lesion with lobulated margins and heterogeneously hyperechoic echotexture overlying the Adductor Pollicis Brevis (APB) muscle. The color Doppler showed increased marginal blood flow with evidence of low-flow internal vascularity suggestive of vascular anomaly.

Multi-planar MRI showed a well-defined soft tissue mass in the thenar eminence of the right hand deeply bounded by the APB muscle, displaying homogenous isointense signal to the muscle on T1 weighted images with bright signal in T1 short tau inversion recovery (STIR) images with signal void vessels inside the lesion suggestive of vascular anomaly (Figs. 2 and 3).

Clinically, in contrast to radiological suggestions, it was clearly a tumor of non-vascular origin.

Total Excision under tourniquet and Loupe magnification was carried out, with direct visualization and preservation of the recurrent motor branch of the median nerve (Fig. 4).

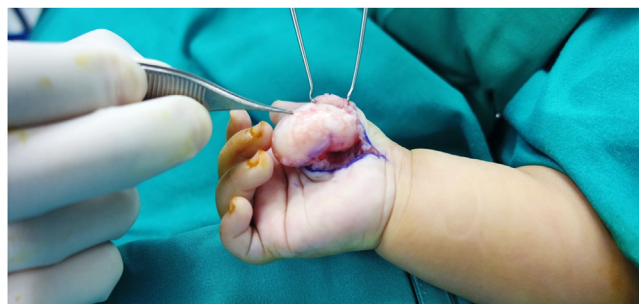


Fig. 4. Intraoperative image of the Nerve Sheath Myxoma.

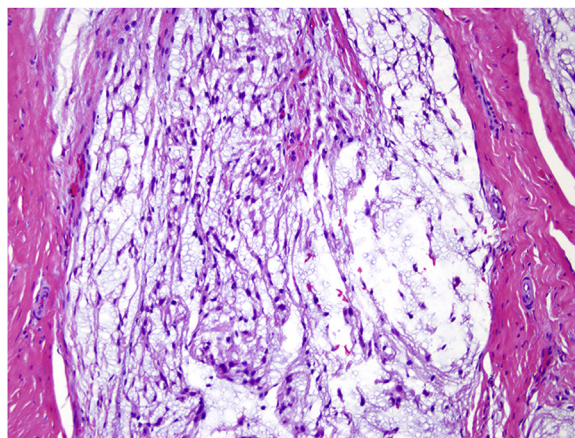


Fig. 5. Lobules containing myxoid matrix separated by collagenous septa.



Fig. 6. Picture showing result 4 months postoperatively.

Histopathology revealed lobules containing myxoid matrix separated by collagenous septa and surrounded by a thick collagenous capsule-like fibrous band. The myxoid nodules contain stellate cells, strongly positive for S100 protein as well as CD34 positive fibroblasts. The histological and immunohistochemical features were consistent with Nerve Sheath Myxoma (Fig. 5).

Follow up of the patient 6 months postoperatively was uneventful, showing full range of movement of the thumb and no evidence of recurrence so far (Fig. 6).

4. Discussion

This case report highlights the difference between Nerve Sheath Myxoma (NSM) and Neurothekeoma (NT), discussing the imaging techniques and their accuracy in diagnosis, and the importance of

managing these cases by an experienced hand surgeon with special interest in paediatric hand tumors.

Benign Nerve Sheath Myxoma is a rare neuro-ectodermal tumor first described 1969 [3] while, Neurothekeoma (NT) is a cellular variant was named afterwards by Gallager and Helwig [4] in 1980. Later, the terms Neurothekeoma (NT) and NSM were interchangeable [2,6]. In 2011, Sheth et al., clearly differentiated between Neurothekeoma and Nerve Sheath Myxoma based on the genetic expression of the cells using microarray analysis [5]. Nowadays, the consensus is that both, NT and NSM are two separate entities [1,2,6,7].

NSM occurs more commonly in young adults with no gender predilection. The peak incidence is between 30–40 years of age [8]. The tumor is most frequently located in the upper limb and rarely affects the head and neck [9]. NT presents in young women twice as common as men, appearing more commonly on the head and neck and rarely in the hands [1,10]. The peak incidence of NSM is the fourth decade, while NT can affect females from infancy to old age with a peak incidence in the twenties [1,2,11].

Histologically, Neurothekeoma was thought to have 3 distinct subtypes: myxoid, cellular and mixed types, depending on the cellularity and amount of matrix [8,11]. Nerve Sheath Myxoma was previously considered as the myxoid type of Neurothekeoma (NT) [12,13].

Microscopically, NSM as described above has a well-circumscribed, multi-lobulated growth pattern with loosely arranged spindle and stellate cells with S-100 protein positivity lying in copious stromal mucin and partitioned by collagen fibers [2]. On the other hand, NT has characteristic epithelioid cells which are not immunoreactive to S-100 protein [1,2,13]. This characteristic feature of NSM, lead to the agreement that it originates from Schwann cells, while the cellular origin of NT is still debatable [1,2,13].

Because many other soft tissue lesions in the hand such as schwannomas [14] and hemangiomas share similar features on Ultrasound and MRI, it is difficult to reach a diagnosis using standard imaging techniques [15]. On MRI both hemangiomas and schwannomas appear isointense to the surrounding muscles in T1 weighted images and hyper-intense in short tau inversion recovery (STIR) sequences [15,16].

Total excision and biopsy remain the standard of care in MSN, with relatively higher rate of recurrence, compared to NT, especially in cases of incomplete excision [1,2,8,9].

Almost 300 cases of NSM and NT are mentioned in literature [17]. Our reported case: 4-month-old girl is by far the youngest reported case of NSM in the literature.

Although, Nerve sheath myxoma and Neurothekeoma are rare benign neoplasms of the peripheral nervous system, they should be considered for the differential diagnosis for localized asymptomatic soft tissue hand swellings in the pediatric group (i.e. schwannomas, neurofibromas, hemangiomas).

Standard imaging techniques are not helpful in diagnosis of such a lesion. Complete excision remains the main stay of treatment so far with relatively low rate of recurrence. Histo-immunochemistry is the only accurate diagnostic tool up to date.

Declaration of Competing Interest

We disclose that we have received no financial and have no personal relationships with other people or organisations that could inappropriately influence (bias) their work

Funding

We received no funding from any source for this work

Ethical approval

This is a case report. N/A. This a case report with no experimental treatment used, and it is exempt from ethical approval by the institution.

Consent

We confirm that Written informed consent was obtained from the parents for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Haitham Khashaba: Study Concept, Data Collection, literature search, writing the paper.

Eman Hafez: Interpretation of the imaging.

Hisham Burezq: study concept and supervision.

Registration of research studies

1. Name of the registry: N/A. this is not a research study
2. Unique identifying number or registration ID: N/A. this is not a research study
3. Hyperlink to your specific registration (must be publicly accessible and will be checked):

Guarantor

Hisham Burezq.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Acknowledgement

We acknowledge the great help by Dr. Rana Al-Abdulghani for the images and the histopathological report of the case.

References

- [1] H. Elwood, J. Taube, Dermal and subcutaneous plexiform soft tissue neoplasm, *Surg Pathol. Clin.* 4 (2011) 819–843.
- [2] A.M. Cimino-Mathews, Peripheral nerve sheath tumors, *Surgical Pathol.* 4 (2011) 761–782.
- [3] J.C. Harkin, R.J. Reed, Tumors of the peripheral nervous system, in: *Atlas of Tumor Pathology*, 2nd ed., AFIP, Washington DC, 1969, 60.
- [4] R.L. Gallager, E.B. Helwig, Neurothekeoma—a benign cutaneous tumor of neural origin, *Am. J. Clin. Pathol.* 74 (1980) 759–764.
- [5] S. Sheth, X. Li, S. Binder, S.M. Dry, Differential gene expression profiles of neurothekeomas and nerve sheath myxomas by microarray analysis, *Mod. Pathol.* 24 (2011) 343–354.
- [6] K.M. Benbenisty, A. Andea, J. Metcalf, et al., Atypical cellular neurothekeoma treated with Mohs micrographic surgery, *Dermatol. Surg.* 32 (2006) 582–587.
- [7] J.F. Fetsch, W.B. Laskin, M. Miettinen, Nerve sheath myxoma: a clinicopathologic analysis of 57 morphologically distinctive, S-100 protein and GFAP-positive myxoid peripheral nerve sheath tumors with a predilection for the extremities and a high local recurrence rate, *Am. J. Surg. Pathol.* 29 (2005) 1615–1624.
- [8] I. Ahmed, J. Rawat, S. Singh, A. Sharma, A. Pandey, M. Goel, Neurothekeoma: a sacrococcygeal tumor in a child, *J. Pediatr. Surg.* 45 (2010) 1037–1039.
- [9] S.A. Fanto, E. Fanto, Neurothekeoma of the median nerve: case report, *J. Hand Surg.* 37A (2012) 1184–1186.
- [10] J.L. Hornick, C.D. Fletcher, Cellular neurothekeoma: detailed characterization in a series of 133 cases, *Am. J. Surg. Pathol.* 31 (2007) 329–340.
- [11] P.W. Allen, Myxoma is not a single entity: a review of the concept of myxoma, *Ann. Diagn. Pathol.* 4 (2000) 99–123.
- [12] S. Uygur, S. Ayhan, S. Kandal, M. Gureli, M. Uluoglu, Giant neurothekeoma in the forearm of an infant, *JPRAS* 62 (2009) 259–262.

- [13] J.F. Fetsch, W.B. Laskin, J.R. Hallman, G.P. Lupton, M. Miettinen, Neurothekeoma: an analysis of 178 tumors with detailed immunohistochemical data and long-term patient follow-up information, *Am. J. Surg. Pathol.* 31 (2007) 1103–1114.
- [14] H. Yamazaki, A. Kaneko, Y. Ota, K. Tsukinoki, Schwannoma of the mental nerve: usefulness of preoperative imaging: a case report, *Oral Surg. Oral Med. Oral Pathol. Oral Radiol. Endod.* 97 (2004) 122–126.
- [15] J. Teh, G. Whiteley, MRI of soft tissue masses of the hand and wrist, *Brit. J. Rad.* 80 (2007) 47–63.
- [16] C.L. Forthman, P.E. Plazar, Nerve tumors of the hand and upper extremities, *Hand Clin.* 20 (2004) 233–242.
- [17] D.J. Koumanis, L.T. Glickman, Facial neurothekeoma in a 10-year-old child, *Can. J. Plast. Surg.* 15 (2007) 175–177.
- [18] R.A. Agha, M.R. Borrelli, R. Farwana, K. Koshy, A. Fowler, D.P. Orgill, For the SCARE Group, The SCARE 2018 Statement: Updating Consensus Surgical Case Report (SCARE) Guidelines, *Int. J. Surg.* 60 (2018) 132–136.

Open Access

This article is published Open Access at [sciencedirect.com](https://www.sciencedirect.com). It is distributed under the [IJSCR Supplemental terms and conditions](#), which permits unrestricted non commercial use, distribution, and reproduction in any medium, provided the original authors and source are credited.