JOURNAL OF CLINICAL AND HEALTH SCIENCES

CASE REPORT

Fibromatosis Colli, A Forgotten Entity

Muhammad Raziin Zainal Abidin[1], Haziq Hakimi Mohamad Azmi[2] , Syarifah Nafisah Syed Hamzah[1], Norazila Abdul Rahim[1,3]

1 Department of Otorhinolaryngology Head & Neck Surgery, Hospital Al Sultan Abdullah, Universiti Teknologi MARA, Puncak Alam, Selangor
2 Department of Otorhinolaryngology, Hospital Cyberjaya, Selangor
3 Department of Otorhinolaryngology Head & Neck Surgery, Faculty of Medicine, Universiti Teknologi MARA, Sungai Buloh, Selangor

Received
13th November 2023
Received in final revised form
28th December 2023
Accepted
13th February 2024
Published
1st March 2025

Corresponding author:
Norazila Abdul Rahim,
Department of Otorhinolaryngology Head & Neck Surgery,
Faculty of Medicine,
Universiti Teknologi MARA, Sungai Buloh, Selangor, Malaysia
Email: norazila3587@uitm.edu.my
Tel. no: +60174383327

ABSTRACT

Fibromatosis colli or congenital muscle torticollis of infancy is a rare cause of neck swelling that originates from the sternocleidomastoid muscle (SCM). It typically presents as an incidental finding of a firm and painless neck swelling by concerned parents. The cause of this pathology is unknown. Ultrasonography (US) is the modality of choice to confirm diagnosis and treatment is mainly by physiotherapy. However, in inexperienced doctors' hands, this pathology may be over investigated with unnecessary minimally invasive techniques such as fine needle aspiration or biopsy to obtain diagnosis. We report a case of a one-month-old boy presented with a painless right neck swelling, management, and literature review with the objective to remind the fraternity of this forgotten diagnosis and to prevent over investigation of this benign entity.

KEYWORDS: Fibromatosis Colli, Sternocleidomastoid, torticollis, head and neck, infant neck mass

INTRODUCTION

Fibromatosis colli was firstly reported as 'congenital muscle torticollis'. It was thought to be caused by formation of hematoma due to injury to SCM [1]. It typically presents as a firm, painless neck swelling which occurs within 2 to 4 weeks of life [2]. The presentation is sudden or incidental finding without any history of trauma. The exact pathophysiology is not well understood. Ultrasonography is the modality of choice in confirming the diagnosis due to high sensitivity [3]. We present a case of a one-month-old boy presented with a painless right neck swelling and management.

CASE PRESENTATION

We present a case of a one-month-old boy who was incidentally found to have a right neck swelling noted by his mother at 23 days of life. The mass was static in size and did not cause any irritability to the child. He had no fever, recent upper respiratory tract infections or any symptoms of inflammation. There were no upper aerodigestive obstructive symptoms. There was no prior history of trauma to the neck. He was born via spontaneous vaginal delivery without any complications or needing any assistive tools. He had no contact with individuals who had tuberculosis.

Clinical examination revealed a firm right neck mass measuring 3 x 2cm fixed along the middle third of the left SCM (Figure 1). The mass was non tender and had no inflammatory skin changes such as punctum or erythema. There was no limited neck range of motion noted. There were no adjacent skin or scalp lesions. The flexible nasopharynglaryngoscope was unremarkable and laboratory markers were within normal ranges.

Ultrasonography (USG) of the neck revealed a bulky and fusiform right SCM (anteroposterior

diameter 1.2cm) compared to the left (AP diameter 0.4cm) (Figure 2). Normal muscle striations are not visualized over the right SCM. There are ill-defined hypoechoic areas within the right SCM. There is no increase in Doppler signal.

A diagnosis of Fibromatosis Colli was made by clinical examination and confirmed by USG findings. The boy was referred to physiotherapy for stretching exercises. No medical or surgical treatment was prescribed. After 6 weeks of physiotherapy, the mass resolved completely.

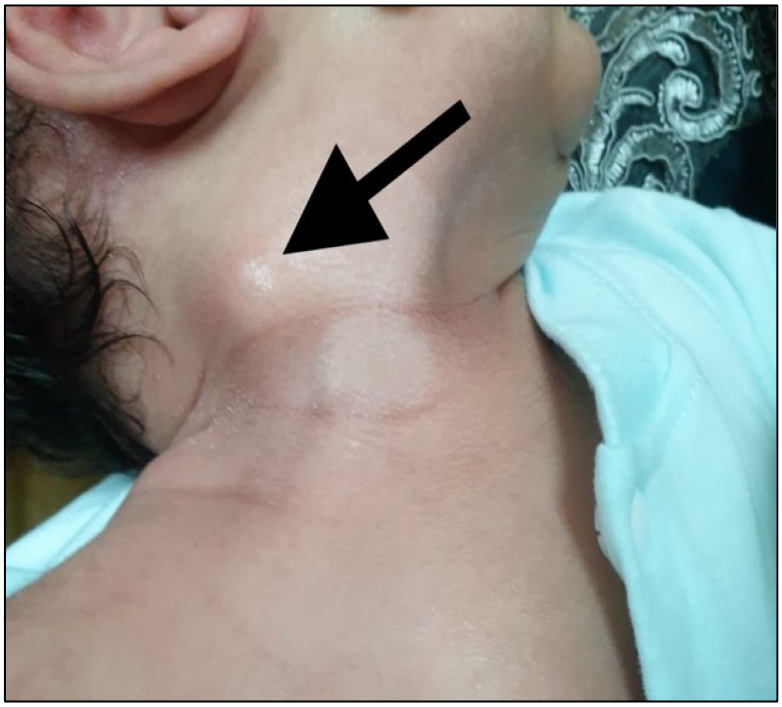

Figure 1 Right neck mass on examination noted by black arrow

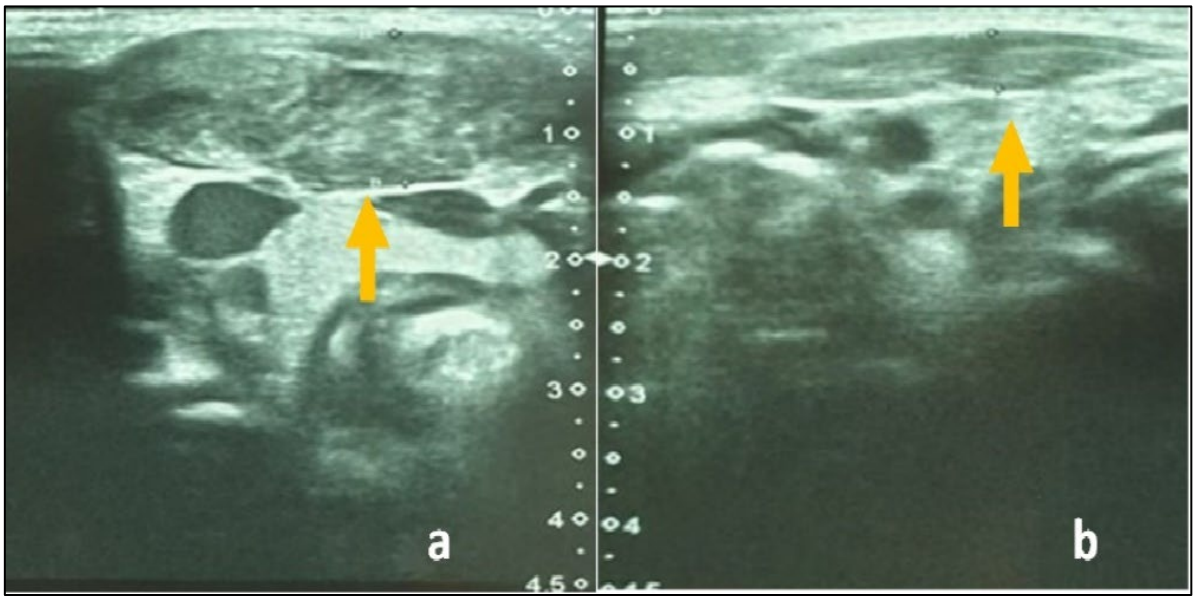

Figure 2 USG neck showed bulky and fusiform enlargement of (a) Right SCM (arrow) when compared to (b) Left SCM (arrow)

Vol 10(1) (2025) 77-81 | jchs-medicine.uitm.edu.my | eISSN 0127-984X
https://doi.org/10.24191/jchs.v10i1.5431

DISCUSSION

SCM is a paired muscle of the anterolateral part of the neck. It divides the neck into anterior and posterior triangles. It consists of two heads (sternal head and clavicular head) and insert from the lateral process of the mastoid process manubrium of sternum and medial third of clavicle respectively [4]. Fibromatosis colli or SCM tumor of infancy is a rare benign cause of neck mass arising from the SCM. Its prevalence is 0.4 %. It occurs most commonly on the right SCM (73%). It is usually detected during 2 to 4 weeks of life and is noted to affect boys more than girls [5]. Patients usually presents with incidental finding of painless neck swelling. Although not present in our case, 20% of cases is reported with torticollis [6].

Fibromatosis colli was first reported by K. F Hulbert in 1965. It was previously reported as congenital muscle torticollis [1]. It was previously postulated that birth injury results in the formation of hematoma within the SCM muscle. However, according to Hubert et al, examination of the tumor revealed no hematoma [1]. The current working hypothesis stipulates that fibromatosis colli develops due to venous obstruction. The impaired venous outflow in the SCM lead to fibrosis of the muscle [2]. Another hypothesis by David et al stated that head positioning in utero can selectively injure the SCM which lead to intra uterine or perinatal compartment syndrome resulting in congenital muscular torticollis [7].

Diagnostic US of the neck is the investigation of choice with 95% sensitivity. It is noninvasive, cheap and may spare the patient from unnecessary radiation [8]. The diagnosis by US is confirmed by showing fusiform enlargement and hypoechoic lesion arising from the SCM [3]. However, if further imaging is required, computed tomography (CT) can be performed which the features of fibromatosis colli will appears as an isodense enlargement of the SCM [3]. As compared to rhabdomyosarcoma, US generally presents as a well-defined, slightly hypoechoic inhomogeneous mass that can show increased flow [12]. Cervical tuberculosis on the contrary will revealed avascularity, displaced hilar vessels, and low intranodal vascular resistance [13]. This case illustrates the importance of recognition of clinical and radiographic findings of fibromatosis colli. The diagnosis was made confidently by clinical examination and confirmed with radiological findings, thus preventing the patient from being subjected to potentially invasive procedures. Fine needle aspiration and cytology (FNAC) has been documented to diagnose fibromatosis colli and rule out potentially malignant diagnosis such as embryonal rhabdomyosarcoma, lymphoma, and neuroblastoma. The microscopic results may easily identify the lesion as benign. The cellular composition comprised of spindle and plump fibroblasts, frequently arranged in loose clusters along with interconnected strands of collagen. On occasions, the fibroblasts appeared devoid of cytoplasm, revealing "naked" ovoid nuclei, resembling a myoepithelial cell origin. The presence of degenerate skeletal muscle was observed, displaying diverse patterns ranging from identifiable myofibers with cross striations to multinucleated giant cells [9]. However, the authors believe that this method maybe spared in cases with clear cut clinical features without any suspicion of malignancy. However, if any suspicion of other pathology following US, FNAC should be considered.

Fibromatosis colli may be treated by both conservative and surgical treatments. Mainstay of treatment for fibromatosis colli is physiotherapy. Studies have shown 90% of cases resolved with physiotherapy alone [7]. If treatment is initiated within 4 months of diagnosis, resolution is expected between 3 to 4 months of initiation [5, 6]. As illustrated in our case, the patient recovered after 6 weeks by mainly undergoing physiotherapy. However, for refractory cases that do not resolve after 1 year of conservative management, patients can be offered surgical tenotomy or Botulinum toxin [10]. In the case if left untreated, they will be deficits in lateral and rotational range of motion and can occur along with irreversible facial and skeletal deformities that develop over time [11].

CONCLUSION

Fibromatosis colli is a rare cause of neck swelling in infancy. The diagnosis can be made by proper clinical examination and confirmation by US which shows fusiform enlargement and hypoechoic features. Besides of 95% sensitivity, it is cheap and noninvasive, sparing

Vol 10(1) (2025) 77-81 | jchs-medicine.uitm.edu.my | eISSN 0127-984X
https://doi.org/10.24191/jchs.v10i1.5431

the patient from unnecessary invasive procedure such as FNAC. Early diagnosis and treatment will lead to early resolution without any complication. We would like to reiterate this in order to achieve diagnosis as it can prevent patients from undergoing unnecessary procedures or invasive methods. The mainstay of treatment is physiotherapy.

Conflict of Interest

Authors declare none.

Acknowledgement

The authors wish to extend their heartfelt thanks to all individuals and organizations who contributed to the success of this research.

We are profoundly grateful to Professor Dr Masaany Mansor for her invaluable guidance, encouragement, and insightful suggestions throughout the course of writing this manuscript.

Our appreciation also goes to our Radiology Department colleague from Hospital Tuanku Ampuan Najihah for their collaborative spirit and constructive feedback, which greatly enhanced the quality of this work.

Additionally, we express our gratitude to the parents of the said patient for their willingness to contribute to this manuscript

Lastly, we wish to thank our families and loved ones for their understanding, patience, and encouragement throughout this journey.

Authors' contribution

- Muhammad Raziin Zainal Abidin: Conceptualized and designed the study, developed the methodology, and wrote the initial draft of the manuscript.
- Haziq Hakimi Mohamad Azmi: Conducted data collection, performed the formal analysis, and contributed to the interpretation of results.
- Norazila Abdul Rahim: Provided critical revisions to the manuscript, validated the data, and supervised the research process.
- Syarifah Nafisah Syed Hamzah: Assisted with the literature review, supported data visualization, and contributed to the final editing and proofreading of the manuscript.

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Vol 10(1) (2025) 77-81 | jchs-medicine.uitm.edu.my | eISSN 0127-984X
https://doi.org/10.24191/jchs.v10i1.5431