

Case Report

A neonate with neck swelling: Fibromatosis colli Boyunda şişlik olan bir yenidoğan: Fibromatozis kolli

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ABSTRACT

Fibromatosis colli is a rare, benign, cervical fibrous tumor observed in neonates and called sternocleidomastoid tumor of infancy. It typically presents as a palpable mass in the anterior part of the sternocleidomastoid muscle that is not present at birth but appears within the first weeks of life. Its etiology remains poorly understood, and most of the patients have a history of abnormal fetal head position and/or birth trauma. In this case study, we report a case of fibromatosis colli, diagnosed using ultrasonography and treated with physiotherapy. We emphasize that we should be aware of this clinical entity in the differential diagnosis of neck mass in neonates to prevent unnecessary investigations and refer them to physiotherapy.

Keywords: Neonate, fibromatosis colli, sternocleidomastoid tumor, ultrasonography

ÖZET

Fibromatoziskolli, nadir olan, süt çocukluğunun sternokleidomastoid tümörü olarak adlandırılan, yenidoğan döneminin benign servikal fibröz tümördür. Sternokleidomastoid kasın ön kısmında görülen, doğumda mevcut olmayan, ancak yaşamın ilk haftalarında ortaya çıkan bir kitledir. Etiyolojisi tam olarak anlaşılamamıştır ve hastaların çoğunda anormal fetal baş pozisyonu ve/veya doğum travması öyküsü vardır. Ultrasonografi ile tanısı konulan ve fizyoterapi ile tedavi edilen bir fibromatozis kolli olgusunu sunuyoruz. Yenidoğanlarda boyun kitlelerinin ayırıcı tanısında bu klinik tablonun hatırlanması ile gereksiz tetkiklerin önlenip hastaların fizyoterapiye yönlendirilmesi gerektiğini vurguluyoruz.

Keywords: Yenidoğan, fibromatozis kolli, sternokleidomastoid kas tümörü; ultrasonografi

INTRODUCTION

Fibromatosis colli (FMC) is a rare, benign, cervical mass observed in neonates and is also called congenital muscular torticollis and sternocleidomastoid (SCM) tumor of infancy. It is classified as a fibroblastic proliferation with a benign nature, constituted by spindle-shaped cells of SCM muscle. Its incidence has been reported

as 0.4%. Although its etiology remains poorly understood, it is believed to be a result of underlying muscle injury, and 50% of the patients have history of muscle injury in utero and/or birth trauma (1-5). It is unilateral, generally right-sided, and more common in male infants (6-8). It presents with a mass located on

Received: 31.08.2024 · Accepted: 16.09.2024 · Published: 25.02.2025

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Cite this article as: Özyay HB, Bekirçavuşoğlu AF, Arslan M. A neonate with neck swelling: Fibromatosis colli. Pediatr Acad Case Rep. 2025;4(1):16-9.

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the SCM muscle in the anterior region of the neck that could be associated with facial asymmetry and torticollis. This mass is absent at birth and develops postnatal, typically between the 2nd and 4th weeks. The diagnosis is mainly clinical, and ultrasonography (USG) is used to confirm the diagnosis (1,2,6-9). USG plays a critical role in differentiating this benign condition from other causes of neck masses in neonates, such as rhabdomyosarcoma, neuroblastoma, lymphoma, cystic hygroma and branchial cleft cyst (3,5,7). In this case study, we reported a case in which physical examination and USG revealed the diagnosis of FMC. The patient's parents provided informed consent for publishing this study.

CASE REPORT

A 3-week-old male neonate presented to the outpatient clinic for the evaluation of swelling in the neck. It was noted that the swelling in the neck had occurred in the previous week, and its dimensions had become greater. There was no history of fever, trauma and infection. The antenatal, natal, and postnatal history, as well as family history, were all unremarkable. The baby was born at term through normal spontaneous vaginal delivery as the first child of nonconsanguineous healthy parents with a birth weight of 3000 g. Systemic and neurological examinations were normal. A firm, painless and immobile swelling, with approximately 1.5-2 cm soft tissue mass was observed in the lower portion of the SCM muscle on the left side of the neck. There was a slight deviation of the neck to the right, and neck movements were mildly limited towards the affected side (Figure 1). USG revealed diffusely increased thickness in the muscle and a hypoechoic appearance on the left SCM muscle. Dimensions and echogenicity of the right SCM muscle were normal (Figure 2). The patient was diagnosed with FMC due to typical clinical and imaging findings. The patient was on physiotherapy and the mass showed a gradual resolution. During the next assessment in the 3rd month, it was observed that the mass had shrunk, and the neck movements were free. Physical examination was completely normal at 9 months (Figure 3).

DISCUSSION

FMC is a disease of infancy that is also called pseudotumor of SCM muscle. It is not a cancerous disease, although the word "tumor" is typically used to describe it. This disease is known as a congenital fibrotic pro-

cess, and the word "tumor" means swelling (1,5,7,10). It was first defined by Hubert, and Chandler and Altenberg then determined its characteristics as a firm, immobile, fusiform swelling on the SCM muscle, which begins to form approximately two weeks after birth and may attain the size of a large almond within four weeks of birth (1,11).



Figure 1. Neonate with swelling in the anterolateral aspect of the neck.

Although the pathogenesis of this lesion has not been clarified, it has been reported that it may be associated with dystocia, breech presentation, forceps birth, and normal spontaneous vaginal delivery. The most widely accepted factor in etiology, however, is hindered venous output of the muscle during birth or intrauterine development. This damage leads to necrosis and, subsequently, fibrosis of the muscle fibers, which causes the formation of secondary stress on the muscle (10-12).

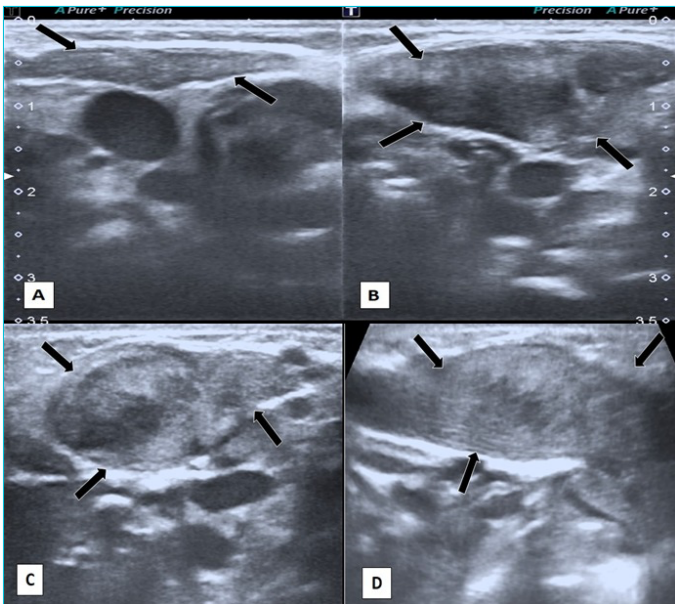


Figure 2. On the left (A), axial ultrasonography (USG) shows a sternocleidomastoid (SCM) muscle of normal size and echogenicity in the anterior jugular vein and main carotid artery. On the right (B), however, a SCM muscle with increased dimensions and hypoechoic, heterogeneous appearance is observed. Axial (C) and longitudinal (D) USG images demonstrate marked thickening and heterogeneous echogenicity changes, suggesting fibromatosis colli on the SCM muscle.



Figure 3. Physical examination appeared normal at 9 months.

Although it is usually unilateral, a bilateral involvement may also be observed rarely. While it is typically seen on the right side, the lesion was left-sided in our patient. Shortening of the SCM muscle occurs following diffuse fusiform growth, leading to rotation of the patient's head to the opposite side of the involved side (5-6). It is the most frequent cause of congenital torticollis, which may lead to facial and cranial asymmetry (2).

FMC should be considered when lateral neck swellings occur in the early term, especially within the first month of birth. Its differential diagnosis includes reactive lymph nodes, abscesses and bronchogenic cysts and malignant conditions, including neuroblastoma, lymphoma, rhabdomyosarcoma, teratoma and fibrosarcoma (3).

USG takes an important place for diagnosis since it is a non-invasive, easy-to-perform, easy-to-access, cost-effective and radiation-free method. Although USG is the imaging method of choice for diagnosis, computed tomography and magnetic resonance imaging are also used for differential diagnosis when suspicious or abnormal clinical findings exist. Surgical intervention and biopsy should be reserved for cases in which imaging studies are equivocal or when the lesion fails to resolve spontaneously (5,7,10).

For our patient, the definitive diagnosis was possible and was made solely using USG; therefore, additional imaging methods were not required.

In general, FMC is self-limited, and treatment is usually conservative with physiotherapy that should begin early to ensure a better outcome. Although the mass regresses spontaneously during the next four to six months, in fewer than 10% of the cases, alternative therapies, including botulinum toxin type A or different surgical methods such as SCM excision, the release of the SCM, tenotomy, may be required (13-15). The prognosis of children diagnosed and treated at older age is worse (7). In our patient, the mass resolved gradually with physiotherapy, and no additional intervention was required.

In most studies, FCM spontaneously regressed with physiotherapy in the first year of life, and no additional treatment was required (5,8,11). In a study conducted with 26 patients, the patients were followed without physiotherapy, and satisfactory results were obtained at the end of the 1st year (10). The same result was reported by Adamoli (15). However, these authors still emphasized the importance of physiotherapy in preventing the development of torticollis.

In conclusion, FMC is a rare, self-limiting, benign pseudotumor of the SCM. The diagnosis is principally made by clinical and radiological findings. The condition should be identified quickly in the neck masses of neonates to avoid unnecessary investigations and promptly begin conservative treatment using physiotherapy for better outcomes.

Patient Consent Form / Hasta Onam Formu

The parents' of this patient consent was obtained for this study.

Conflict of Interest / Çıkar Çatışması

The authors declared no conflicts of interest with respect to authorship and/or publication of the article.

Financial Disclosure / Finansal Destek

The authors received no financial support for the research and/or publication of this article.

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