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Fibromatosis Colli in Neonate

Medical Science

KEYWORDS : Sternocleidomastoid, pseudotumor of infancy, fibromatosis Colli, vacuum extraction, forceps delivery, tenomyotomy

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ABSTRACT

Fibromatosis colli occurs in sternocleidomastoid muscle with a slight male predilection which is common cause of infantile torticollis. Predominantly it presents within first weeks of life. It is mostly related to birth trauma due to breech, forceps delivery. Fibromatosis colli is to be recognized early as benign lesion that it is. It is unilateral in most cases. Imaging wise- modality of choice is ultrasonography. Mostly it's a self-limiting condition resolves mostly with physiotherapy. Rarely requires surgical correction if treatment is delayed due to delayed diagnosis.

Introduction

Painless diffuse enlargement of the sternocleidomastoid muscle also called as Fibromatosis colli is a frequently seen in infancy [1]. The exact pathogenesis is unknown, it is mostly due to birth trauma [2]. Various theories related for cause of Sternocleidomastoid impairment in CMT include intrauterine crowding [3-4], muscle trauma during a difficult delivery, [5-7] soft tissue compression leading to compartment syndrome, [8] and congenital abnormalities of soft tissue differentiation within the SCM muscle.

It is one of the causes of congenital torticollis. Imaging modality of choice is USG but cross sectional imaging with CT scan or MRI may be required to further characterize the disease and to know the extent of involvement. We present a case report where fibromatosis colli was primarily diagnosed using MRI, in an infant.

Discussion

A 23 days old female neonate was referred to the radiology department for MRI of brachial plexus with significant birth history; baby born with full term breech extraction with forceps with fracture of left clavicle. Parent noticed restriction of upper limbs and neck movements since 1 week and also baby did not cried immediately after birth. On local examination the right sternocleidomastoid was mildly bulky which is firm to hard in consistency, no e/o erythema edema, or local raised temperature. No h/o fever.

MRI showed the most of the right sternocleidomastoid muscle appears bulky with T2 heterogeneously hyperintense (Figure 1 coronal and axial T2 WI images) with few hypointense signal within, showing no blooming on GRE and no restriction of diffusion on DWI. No e/o T1 hyperintense components within (fat) (Figure 2 coronal and axial T1 WI images). There is resultant mild torticollis to the right.

Left sternocleidomastoid muscle appears normal.

There was no cervical lymphadenopathy. Retrospectively USG of bilateral neck is done. It revealed a thickening of right sternocleidomastoid muscle; with fusiform appearance and heterogenous echotexture (Figure 3). However fibrillary structure of muscle fibers was maintained. On colour Doppler image of bulky right Sternocleidomastoid muscle showing no significant vascularity within (Figure 5).In

comparison, the left sternocleidomastoid muscle appeared normal (Figure 4).

Based on these MRI, USG features, history and the clinical findings, a diagnosis of fibromatosis colli or pseudo tumor of the sternocleidomastoid muscle was considered. Physiotherapy was started.

Most commonly the cases of fibromatosis colli also called as pseudotumor of the Sternocleidomastoid presents at 2-4 weeks of birth with a neck swelling, predominantly after difficult delivery (vacuum extraction or forceps delivery).

Pseudotumor and congenital muscular torticollis probably represent different manifestations of SCM muscle fibrosis. Fibromatosis colli was first described as "Sternomastoid tumor torticollis" in German literature in 1812 by Hulbert [11]. It was characterized by Chandler and Altenberg [3] as the appearance of "a hard, immobile, fusiform swelling in the sternomastoid muscle which usually is detected at 2 weeks after birth and then increases in size for two to four weeks until it reaches the size of a very large almond." The mean ages of presentation is 24 days.

They are usually common unilaterally but rarely found bilaterally. Fibromatosis Colli is more common on the right (73%–75%) than the left side of the neck (22%) [4, 8]. A consistent relationship between congenital muscular torticollis and dysplasia of the hip is widely accepted. Primary diagnosis was made on USG, which reveals spindle-shaped thickening of the sternocleidomastoid muscle of the affected muscle as in compared to the normal contralateral side. The ultrasound can distinguish the arrangement and composition of muscle fibers [10]. Type I fibrosis showed a localization, and type II (a diffuse type) was shown by a mixture of fibrous tissue and nor-

mal muscle fibers in the involved SCM. Type I and type II fibrosis may represent a transitional state and develop types III and IV, which represent an irreversible state of CMT in which there is severe fibrosis with nearly complete absence of normal muscle tissue. Types III and IV fibrosis initially may contribute to the persistent symptoms up to advanced age and the high incidence of surgical intervention. Consequently, it is possible that if the patient is a type I or II,

conservative treatment may be adequate, while type IV patients may require a surgical approach. There is no cervical lymphadenopathy and no vascular invasion or bony involvement as may be seen with other neoplastic lesions. The mass increases in size until one month of age, remains static for two to three months,

then gradually diminishes in size and disappears clinically [9-10]. However, the resulting fibrosis of the sternocleidomastoid muscle may lead to congenital muscular torticollis in 10 to 20 percent of cases [12]. Sometimes bilateral sternocleidomastoid tumors of infancy, though extremely rare, have also been described (13). Treatment is symptomatic, with physiotherapy and neck stretching exercises. Tight bands of residual fibrosis resulting from contracture of the SCM muscle could be present after resolution of the pseudotumor in children for whom conservative intervention does not yield satisfactory results. Botulinum toxin (Botox) could enhance the effectiveness of stretching on the side of the contracture and allow strengthening of overstretched and weakened muscles on the opposite side of the neck. Surgical treatment like tenomyotomy or tenotomy is required and is indicated when a patient has undergone at least six months of controlled manual stretching and has residual head tilt; deficits of passive rotation; lateral bending of the neck less than 15 degrees; and a tight muscular band or tumor. Surgery usually is not considered for those under one year of age [14]. Possible differential diagnosis of solid tumors in this age group includes rhabdomyosarcoma and neuroblastoma in this along with neck mass, there can be associated enlarged cervical lymph nodes, vascular involvement, or invasion of surrounding structures. Neck reactive lymph nodes, abscesses, bronchogenic cysts and thyroid lobe dislocation are clinically easily distinguished from fibromatosis as well as malignant conditions such as Neuroblastoma, lymphoma, rhabdomyosarcoma, teratoma and fibro Sarcoma [10] and benign neoplasmatic conditions such as hemangioma and cystic

hygroma. In the differential diagnosis also vascular malformation (particularly lymphatic malformation), beside bronchogenic cysts and thyroglossal duct cysts, is to be evaluated.

USG is the modality of choice to identify the lesions [1]. On CT scan, the sternocleidomastoid muscle appears diffusely enlarged, isodense in attenuation [1].

MRI features include decreased signal intensity of the mass on T2W images as compared to gradient-recalled T1W images, because of the presence of fibrous tissue [15]. The further characterization of the disease and extent of involved muscle is better delineated with MRI and CT.

To conclude, fibromatosis colli less common cause of neck swelling neonates and infants and the radiologist must be aware of its imaging features in order to early diagnosis and to differentiate it from other neck masses.

Figure 1-- coronal and axial T2 WI images showing bulky right sternocleidomastoid muscle with T2 heterogeneously hyperintense with few T1 hypointense signal within

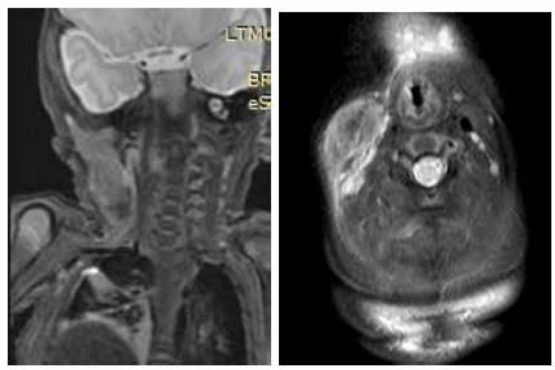

Figure 2 -- coronal and axial T1 WI images showing bulky hypointense right sternocleidomastoid muscle with no T1 hyperintense components within (fat)

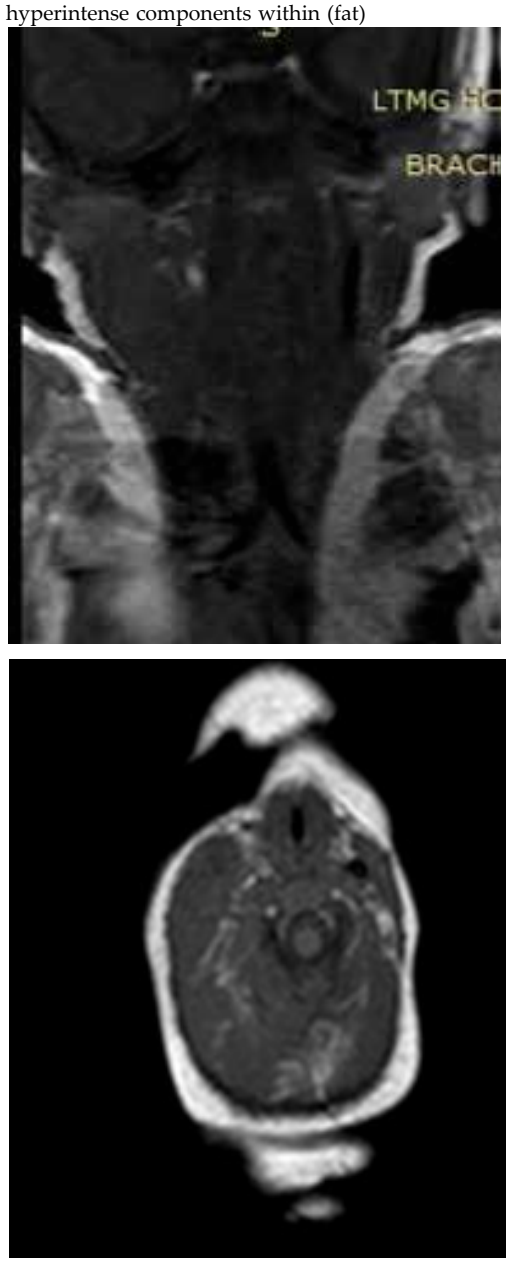

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Figure 3 --gray scale US image of bulky right sternocleidomastoid muscle

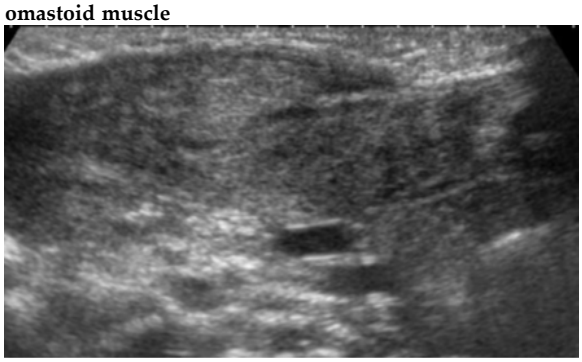


Figure 4 gray scale image of normal sternocleidomastoid muscle

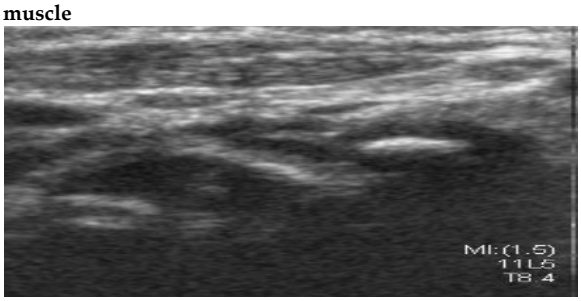
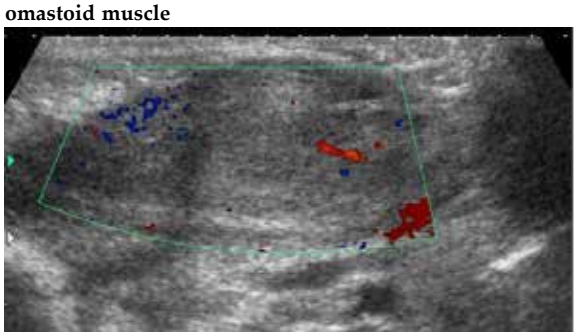


Figure 5 colour Doppler image of bulky right sternocleidomastoid muscle



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