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FIBROMATOSIS COLLI OF INFANCY – THE ROLE OF ULTRASONOGRAPHY : A CASE REPORT

KEYWORDS

Fibromatosis colli, pseudotumour of sternocleidomastoid, ultrasonography, infant

DR MASHOO NEIL DUBEY

ABSTRACT Fibromatosis colli or sternocleidomastoid tumor of infancy is a condition of benign proliferation of fibrous tissue within the sternocleidomastoid muscle leading to focal or diffuse enlargement of the muscle and is often clinically associated with torticollis. Radiological imaging especially ultrasound, if performed by an expert radiologist plays an important role in differentiating this benign condition from other causes of neck masses in this age group, thereby preventing unnecessary investigations in a neonate and decreasing parent`s anxiety. We hereby, present a case report of a sternomastoid pseudotumor in a nine week old neonate diagnosed using high frequency ultrasound, signifying that every physician should be aware of this clinical entity in a neonate and thus prevent irrelevant diagnostic and therapeutic maneuvers.

INTRODUCTION

Fibromatosis colli also known as "sternocleidomastoid pseudotumor of infancy," is an uncommon benign lesion of the spindle cells of the sternocleidomastoid (SCM) muscle resulting in diffuse enlargement of the muscle. The term "tumour" is a misnomer because it is not a cancerous condition but it is referred to as a congenital fibrotic process. Though the exact etiology is not known, it is most likely due to birth trauma. This lesion affects infants with an incidence of 0.4%. It is usually unilateral, affects the right side in 75% of cases, and affects male patients slightly more often than female patients. The mass is not present at birth but tends to appear between the 2nd and 4th week of life. It is one of the causes of congenital torticollis. Though USG is the imaging modality of choice, cross sectional imaging with CT scan or MRI may sometimes be required to further characterize the disease and to know the extent of involvement. Real time USG shows synchronous motion of the mass with the sternocleidomastoid muscle, thus confirming the diagnosis. This is a case report where fibromatosis colli was diagnosed using USG, in an infant.

CASE REPORT

A nine week old male infant was referred to the Department of Radiodiagnosis, SDMH, Jaipur, for evaluation of a left sided neck swelling that had been noticed by the parents since three weeks. The mother also noticed restricted neck movement on the affected side with persistent right sided torticollis. The baby was born after forceps delivery at 40 weeks of gestation, with a birthweight of 3.2 kg, height of 49 cm, and head circumference of 35 cm. Family history was negative for fibrous tissue tumors or congenital torticollis. The baby was the first born child of healthy nonconsanguineous parents. The mother denied having had infectious diseases during pregnancy and having taken drugs or alcohol.

On clinical examination, the swelling was apparent and seen in left suprasternal region [Figure 1] .It was firm to hard in consistency and was not warm to touch. . The baby was otherwise normal, feeding at the breast, and the body temperature was normal. The routine laboratory tests were normal.

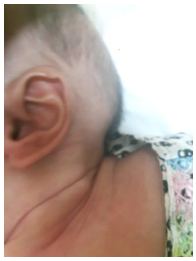

Figure 1. Prominent left sternocleidomastoid in mid distal thirds, in an infant with torticollis

USG neck was performed which documented a thyroid gland of normal size and echostructure with thickened left sternocleidomastoid muscle in lower two third, upto its sternal attachment with heterogenous echotexture [Figure 2].

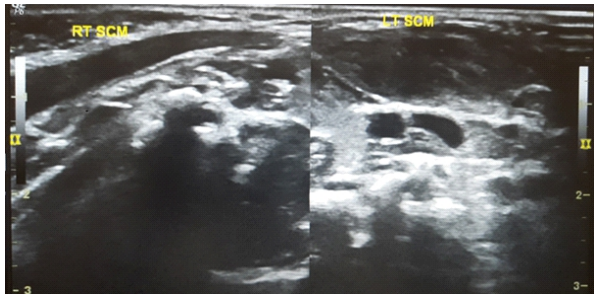

Figure 2: Longitudnal scan of neck in an infant shows fusiform swelling of left sternocleidomastoid in distal thirds with normal right sternocledomastoid

The fusiform shaped mass, which moved synchronously with the muscle, appeared predominantly isoechoic to the rest of the muscle. The fibrillar structure of the muscle fibers was however maintained. No evidence of raised vascularity or focal calcifications were seen [Figure 3].

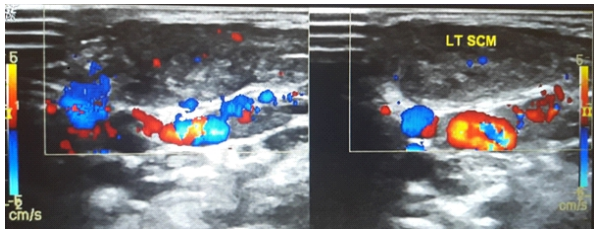

Figure 3: Transverse and longitudinal scan of left sternocleidomastoid muscle shows normal vascularity.

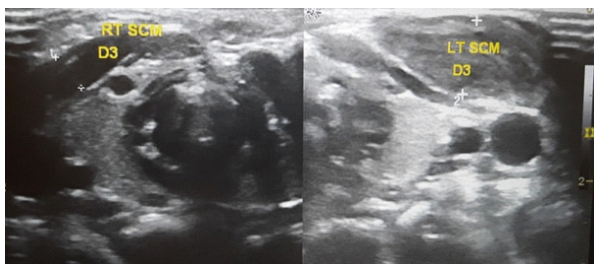

Figure 4: Longitudnal USG scan of neck depicting involvement of left sternocleidomastoid upto its sternal attachment.

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The right sternocleidomastoid muscle appeared normal [Fig 4]. No cervical lymphadenopathy was seen. Rest of the structures of neck were unremarkable. Based on the clinical and USG findings, a diagnosis of fibromatosis colli /pseudotumour of left sternocleidomastoid was considered.

Physiotherapy was started and the swelling gradually reduced with near normal neck movements seen on follow up.

DISCUSSION

Fibromatosis colli was first described as "sternocleidomastoid tumour torticollis" in German literature in 1812 by Hulbert[3]. It was characterized by Chandler and Altenberg[4]. It is a rare form of infantile fibromatosis that occurs within the sternocleidomastoid muscle. It is usually common unilaterally but rarely can be found bilaterally. It is more common on the right (73-75%) than the left side of the neck (22%) [5,6] . There may be a slight male predeliction. It presents a few weeks after birth, then stabilizes in size for few months and finally diminishes spontaneously by 4-8 months of age[7] . Sometimes presentation is with torticollis (seen in 14-20 % of cases)[8,9]. Its pathogenesis is usually related to birth trauma for prolonged labour; breech presentation, forceps delivery or during intrauterine development (malposition in the womb.)[8] . The injury leads to necrosis and fibrosis in the muscle fibres resulting in development of secondary pressure within the muscle. [10-12]. Such an injury leads to a secondary compartment syndrome and resultant pressure necrosis and fibrosis within the muscle. [10,12]

The cytological features include bland appearing fibroblasts and atrophic skeletal muscle, along with muscle giant cells and bare nuclei.

Most authors, in agreement to our findings report USG to be the preferred diagnostic tool because of its easy availability, low cost, and lack of ionizing radiation.[10-12]. USG shows spindle shaped thickening of the muscle with resultant shortening and therefore head turned away from the affected side. Echogenicity may vary. Color Doppler interrogation may reveal high resistance waveform. The affected area often moves synchronously with the rest of the muscle on real time USG [13]. There is no cervical lymphadenopathy and no vascular invasion or bony involvements seen with other neck masses. Torticollis develops in 20%cases. Bilateral sternocleidomastoid tumours of infancy, have also been described.

Cross sectional imaging may occasionally be used to exclude other conditions when the clinical findings are equivocal or atypical and may be required to further characterize the disease and find out the extent of involvement. On CT scan, the muscle appears diffusely enlarged, isodense in attenuation. Adjacent fat planes are well preserved. At times, calcifications may be present[14]. 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. The extent of involved muscle is better delineated with MRI than USG.

Treatment is symptomatic, with physiotherapy and neck stretching exercises. Rarely tenomyotomy or tenotomy is required[15-16]. The swelling regresses over a period of time, with complete disappearance by 4-6 months.

Differential diagnosis includes benign conditions such as hemangiomas, cystic hygroma and malignant conditions like rhabdomyosarcoma, teratoma, lymphoma and neuroblastoma in which, in addition to neck mass, there can be enlarged cervical lymph nodes, vascular encasement or invasion of surrounding structures. Vascular malformation, lymphatic malformations, bronchogenic cysts, thyroglossal duct cysts should be ruled out[11]

CONCLUSION:

Pseudotumour of sternocleidomastoid is a self resolving entity with spontaneous resolution of the lesion over a period of 4 to 8 months. In very young children, the degenerated muscle fibers are replaced with immature fibrous tissue with minimal scarring, as opposed to adults where scar like collagen forms resulting in scarring. USG not only helps in identifying the lesion, but is an excellent modality for real time imaging, where movement of the lesion with respect to the muscle can be identified. It also helps in delineating the extent of the involved muscle and is of great help in follow up of these lesions.

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