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Case Report: Fibromatosis Colli - A Rare Tumor in Infants

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Abstract: ***Fibromatosis Colli also known as congenital torticollis is a rare cause of benign cervical pseudo tumor in neonates, consisting of benign fibrous tissue proliferation within the sternocleidomastoid muscle, resulting in a fusiform enlargement. The origin of fibromatosis colli has been linked to traumatic birth, intrauterine malposition, or vascular occlusion resulting in hemorrhage and, subsequently, fibrosis. . Because of its low cost, lack of ionizing radiation, and the fact that no sedation is needed, Ultrasonography has proven to become the modality of choice when evaluating infants who present with a pediatric neck mass. Ultrasound plays a necessary role in confirming this diagnosis and follow - up. We Report a case of Fibromatosis colli in a six - week old newborn who was diagnosed using Ultrasonography and treated with physiotherapy.***

Keywords: Fibromatosis colli, sternocleidomastoid, infant, neck mass, congenital, pediatric, torticollis

1. Introduction

Fibromatosis colli is a rare condition in which there is diffuse enlargement of the sternocleidomastoid muscle, usually seen in infancy. The worldwide prevalence of fibromatosis colli is estimated at about 4 in every 1, 000 live births. It typically presents as a palpable mass in the anterior neck over the sternocleidomastoid muscle. The mass is typically not present at birth, but it appears within the first few weeks of life, and it may enlarge over the ensuing weeks. It is a focal or diffuse enlargement of the sternocleidomastoid muscle giving an aspect of a cervical pseudo tumor. Though the exact etiology is still unknown, it has been reported that birth trauma or muscle injury in utero could be incriminated in the appearance of this pseudo tumor. The diagnosis is mainly clinical, the unilateral cervical enlargement could be associated with facial asymmetry and limited movement of the neck, called congenital torticollis. However, the ultrasound is able to confirm the diagnosis, eliminate the differentials and ensure the follow - up. This pseudo tumor natural evolution is usually spontaneous in 4 to 8 months requiring only physiotherapy as treatment. We present a case report of congenital torticollis in a six - week - old newborn who had a right cervical swelling that the mother had noticed a few days before.

2. Case Report

A six - week - old newborn (male), was brought with right cervical swelling and restricted neck movement. The obstetrical history of the mother revealed that the pregnancy reached full term, but was poorly monitored, and the newborn was delivered by forceps secondary to prolonged labor. Baby cried immediately after birth, and there was no immediate postnatal complication. The clinical examination revealed a swelling of firm consistency without any inflammatory sings of approximate size 2 cm × 3 cm on the middle portion of the right sternocleidomastoid muscle. It was fixed and non - tender. No other symptom was associated. A cervical ultrasound revealed a fusiform thickening of the right sternocleidomastoid muscle, which was homogeneous in echo - structure, with a preserved muscle's fibrillar structure. No significant change in internal vascularity was seen. This pseudo tumor measured 9.3 mm compared to 4.9 mm on the left side, without mass effect on the vascular axis or the adjacent structures. The thyroid gland on right side of the neck appeared sonographically normal. The sternocleidomastoid muscle on the left side was normal. There was no evidence of hematoma or lymphadenopathy. Based on these findings, we were able to confirm the diagnosis of right sided Fibromatosis Colli. Physiotherapy was started and the swelling showed a slight decrease in size after 3 weeks, with the neck movements returning to near normal. The newborn benefited from motor physiotherapy, we advised his parents to gently rotate the head of the child toward the side of the lesion, five - to six times daily and hold it for a few seconds. At the end of 6 months of physiotherapy the swelling has completely subsided with a near normal ultrasonography picture.

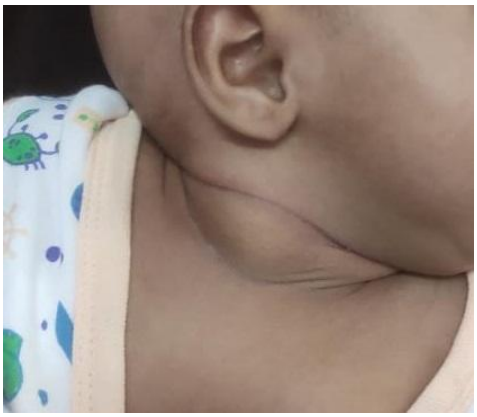

Figure 1: Right side sternocleidomastoid muscle swelling

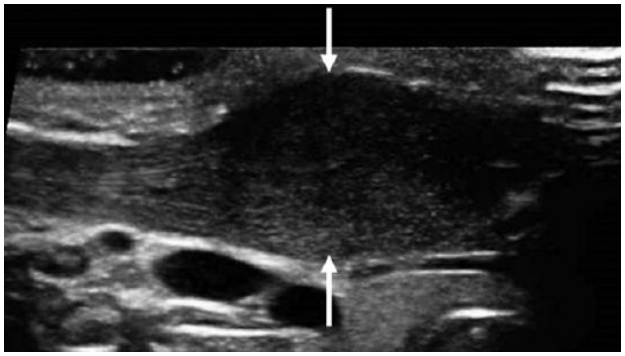

Figure 2: USG picture showing fusiform thickening of the right sternocleidomastoid muscle

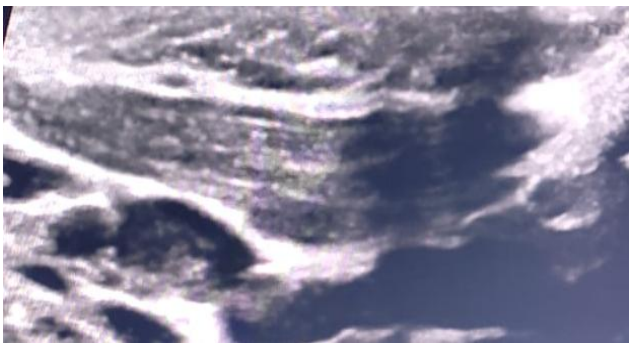

Figure 3: USG picture of right sternocleidomastoid muscle after 6 months of physiotherapy.

3. Discussion

Fibromatosis Colli is a rare condition, with a reported prevalence of 0.4 [2] Fibromatosis colli tumors are characterized by the deposition of benign spindle fibroblasts and collagen around decomposing multinuclear muscle fibers. [3]They are frequently discovered within the first 8 weeks after birth. [3] It is located on the right side in 73 % of cases, and mainly concerns the male gender. [2]The most common presenting manifestation of fibromatosis colli is the presence of a firm, nontender, spindle - shaped nodule, usually in the distal one - third of the sternocleidomastoid muscle. These masses are attached to or located within the body of the sternocleidomastoid muscle, and they are mobile beneath the skin. [3]. Fibromatosis colli lesions attain maximum size within the first month after onset, and most of them gradually regress thereafter, with complete disappearance by 4 - 6 months with some lesions progressing to cause facial and skull deformities. [3, 6]

Grossly, the head of the neonate may be tilted toward the affected side, and there may be a concomitant chin rotation to the contralateral shoulder secondary to sternocleidomastoid muscle contracture. [3]

There is little agreement about the pathophysiology of fibromatosis colli. Difficult delivery has been implicated in the development of these lesions. There are well - established associations between fibromatosis colli and a breech presentation, forceps delivery, and primiparous birth. [7]According to one report, lesions develop during intrauterine growth in approximately 25% of cases, possibly as a result of intrauterine crowding. [8] Fibromatosis colli has been found in newborns following cesarean birth, and there has been an association between fibromatosis colli and several other congenital lesions, including hip dysplasia. [9] One possible cause is venous outflow obstruction in the muscle, either during delivery or during intrauterine development; the working hypothesis is that impaired venous outflow in the sternocleidomastoid muscle can lead to local edema, degeneration of muscle fibers, and ultimately fibrosis of the muscle. The differential diagnosis of pediatric neck masses is extensive and certainly beyond the scope of this discussion. However, fibromatosis colli may be safely diagnosed on the basis of the lesion's natural history and clinical presentation. No other entity presents so soon after birth with the anatomic specificity and physical characteristics of fibromatosis colli. [3]The diagnosis is facilitated by radiographic means. Most recent studies have shown that ultrasonographic examination is sufficiently sensitive and specific c to make the diagnosis. Indeed, Maddalozzo and Goldenberg reported that ultrasonography was 100% sensitive for diagnosis when fibromatosis colli was suspected. [3] The diagnosis can be made on USG, which shows spindle - shaped thickening of the sternocleidomastoid muscle on the affected side in contrast to the normal contralateral side. [1] There is no cervical lymphadenopathy and no vascular invasion or bony involvement as may be seen with other neck masses. More complicated presentations, however, necessitate the use of CT or MRI. On CT scan, the sternocleidomastoid muscle appears diffusely enlarged, isodense in attenuation. MRI features include decreased signal intensity of the mass on T2W images compared to gradient - recalled T1W images, because of the presence of fibrous tissue. The extent of involved muscle is better delineated with MRI than with USG. [6] Fine - needle aspiration may be warranted when other etiologies are suspected. Cytology of fibromatosis colli characteristically reveals mature fibroblasts with variable amounts of striated muscle fibers in different stages of degeneration including bland - appearing fibroblasts and atrophic skeletal muscle, along with muscle giant cells and bare nuclei with no evidence of hemorrhage or inflammation. [4, 5], Nothing relieves the parents more than informing them that the "tumor" their child having is benign or self - limiting. Fibromatosis colli has been managed both conservatively and with surgical intervention. Many studies have shown that 80 to 90% of patients respond to conservative management with physiotherapy. [3] Traditional management has consisted of observation with massage and active and passive stretching. Passive neck stretching exercises allow for strengthening of the affected side, with regression of the mass occurring between four and eight months when progressive physical therapy is used. [3, 2]

Since it is a self - limiting condition reassurance of parents, observation and physiotherapy is all that is required. Surgical intervention can be performed when physical therapy is not successful. However, fewer than 20% of infants who have fibromatosis colli will need surgery for this condition. In terms of surgical intervention, tenomyotomy has been practiced, but it should be reserved for progressive cases and for cases that remain refractory to conservative treatment after 1 year of treatment. [1, 4, 6] Botulinum toxin is currently being explored as an alternative treatment for children with ongoing symptoms despite physiotherapy, but

further evidence of safety and long - term sequelae is required before this is accepted as a suitable treatment. [10]

4. Conclusion

It is recommend that early management of fibromatosis colli include observation and physiotherapy to prevent or reverse torticollis and the craniofacial asymmetry that can result. Conservative management is clearly preferable to any surgical intervention in this group of population, particularly in light of the probability of spontaneous resolution. Similarly attractive is the opportunity that physiotherapy provides for parents to involve themselves in the care of their newborn. A sound knowledge of ultrasound features of fibromatosis colli may obviate the need for relatively unsafe and invasive diagnostic procedures. The value of ultrasound in diagnosing this condition and thereby preventing further unnecessary investigations and uncalled for therapeutic interventions cannot be overemphasized. It is important, therefore, to quickly identify fibromatosis colli in order to avoid any unnecessary expenditures of resources and to promptly begin conservative treatment.

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