Congenital Fibromatosis Colli: A Case Report of rare cause of neck swelling with clinicopathological pointers

Abstract

Fibromatosis Colli or congenital fibromatosis Colli (also known as sternocleidomastoid tumour or pseudotumour) is a rare benign lesion affecting the sternocleidomastoid muscle that manifests in the first few weeks of life and may be associated with muscular torticollis. It is seen in infants following difficult delivery and usually appears as a well-circumscribed, firm to hard, immobile, fusiform swelling in the mid or distal one-third of the sternocleidomastoid muscle. It should be differentiated from more common causes of lateral neck masses including inflammatory, congenital and neoplastic conditions. The disease is diagnosed using typical clinical presentation and fine needle aspiration cytology, however, radiological investigation like ultrasound scan and Computed Tomography scan can help in ruling out other differential diagnosis especially in atypical clinical presentation. We present a rare cause of neck mass that should be kept in mind when evaluating young infants with neck masses.

Keywords: Fibromatosis Colli, sternocleidomastoid tumour, newborn, fine needle aspiration, cytology

Introduction

Fibromatosis Colli (FC) also called congenital fibromatosis Colli or pseudotumour of the sternocleidomastoid muscle is an uncommon benign lesion affecting the sternocleidomastoid muscle that manifests in infancy.[1,2] It typically appears as a diffuse enlargement of the sternocleidomastoid muscle especially in its lower portion during infancy and hence it is called sternocleidomastoid tumour of infancy.[3] It usually presents as a well-circumscribed, firm to hard, immobile, fusiform swelling in the lower or middle portion of the longitudinal anatomic course of the sternocleidomastoid muscle and usually appears within the first few weeks of life.[4] It has an incidence of 0.4%, usually unilateral involving the right side and mostly seen in males.[5] The aetiopathogenesis of the lesion is not fully understood. Various theories proposed include fetal

malposition, birth trauma, ischemic necrosis from vascular compression during a difficult delivery, infection, and the presence of endogenous factors.[6] About 50% of patients with FC have a history of complicated labour, assisted delivery, and breech deliveries.[7,8] Diagnosis can be made based on clinical presentation and investigations like fine-needle aspiration cytology (FNAC) and Ultrasonography, with different researchers and authors having a preference of either of the two.[9,10,11] In most cases, FC resolves spontaneously over time or in response to physiotherapy. Surgical treatment is only required in about 5% of patients that present early but is necessary for about a half of patients presenting after 6 months of age.[12]

Case Report

A 5-week-old male infant presented with painless progressive neck swelling on the right side of the anterior aspect of the neck noticed by the mother two weeks prior to presentation. The mother also noticed preferential tilting of the head towards the right. He was a full-term baby whose labour was complicated by a footling breech presentation. There was no other significant prenatal, natal, or postnatal morbid history. Clinical examination revealed a non-tender swelling, firm to hard in consistency involving the lower and mid-portion of the right sternocleidomastoid muscle measuring 2cm x 3cm and attached to it (Figure 1). The swelling was more prominent with extension movement of the neck. There was a restriction of movement of the neck with tilting of the neck to the ipsilateral side. Other ears, nose, and throat examinations were virtually normal. An ultrasound scan revealed a well-defined fusiform isoechoic mass measuring 4.2cm x 1.9cm which is continuous with the right sternocleidomastoid muscle (Figure 2A & 2B). It is superficial and compressed the adjacent vessels. A Doppler interrogation revealed minimal blood flow within it. Computed tomographic scan showed a well circumscribed fusiform isodense mass involving the mid-lower third of the right sternocleidomastoid muscle compresses the adjacent

internal jugular vein (Figure 3A & 3B). It also showed minimal peripheral enhancement of the mass at the delayed phase of post-contrast images. Fine needle aspiration cytology showed sparsely cellular smear composed of multinucleated muscle cells along with a few bland and spindle cells in small clusters and single in the myxoid background (Figure 4A & B). A diagnosis of the sternocleidomastoid tumour was made and the patient was commenced on physiotherapy with the mass barely noticeable after 3 months of physiotherapy.

Discussion

Fibromatosis colli also called "sternocleidomastoid pseudotumour of infancy" is a self-limiting, benign tumor of infancy, arising from the sternocleidomastoid muscle and presenting shortly after birth.[7] Its aetiology is uncertain, however, various theories have been postulated which include fetal malposition, birth injury, vascular compression causing ischemic necrosis, infections, and the presence of endogenous factors, but it presents most commonly following difficult delivery (vacuum extraction or forceps delivery).[4,6,7] It typically presents with a neck swelling which appears most commonly on the right between 2-8 weeks of life, typically following difficult delivery.[7,13] The index case presented with a right neck swelling noticed at 3 weeks of age following a complicated breech delivery, the swelling has continue to increasing in size since noticed. Fibromatosis Colli naturally initially increases in size, then stabilizes and regresses towards the 4^{th} -8^{th} month of life.[14] Just like the index case, it usually appears as a firm, painless, fusiform mass, 2–3 cm in diameter in the mid or lower portion of the sternocleidomastoid muscle, most commonly on the right, but may be rarely bilateral.[7,13,15] Fibrosis within the lesion can lead to tightening of the muscle with subsequent restriction of movement and ipsilateral tilting of the head refer to as muscular torticollis similar to finding in this report. Torticollis was reported in 10%–20% of cases of FC.[14] In addition,

sternocleidomastoid tumour may be associated with musculoskeletal deformities like ipsilateral mandibular asymmetry, plagiocephaly, facial deformities and postural Cervico-thoracic scoliosis.[14,16] Early diagnosis is essential to differentiate FC from other causes of neck masses in infants including congenital lesions, as well as inflammatory and neoplastic conditions such that appropriate management can be instituted. Non-invasive investigations such as computed tomography (CT) and ultrasonography (USG) can locate the tumour within the SCM, and differentiate solid and cystic lesions. Ultrasonography is usually the initial imaging modality and may show the well-defined non-cystic ovoid or fusiform homogenous mass within the SCM muscle.[14] Some patients may require additional imaging to further evaluate their lesion as seen in this report in which torticollis is present and may be associated with other complications. Furthermore, it can be difficult to differentiate FC from surrounding soft tissue, in such instances, CT scan or Magnetic resonance imaging can be used to distinguish tumour from muscle, fat, or even scar tissue.[14] CT scan typically shows a non-cystic, heterogeneous, isodense mass or enlargement within the boundaries of the sternocleidomastoid muscle.[2] Fine-needle aspiration cytology (FNAC) is minimally invasive, cost-effective, quick, safe, reliable and well-established procedure for the evaluation and diagnosis of head and neck masses FC inclusive. The cytological features include scanty to moderately cellular smears with predominantly slender, spindle to oval shaped cells either singly scattered or clustered and having wispy cytoplasm. There are varying numbers of atrophic degenerated skeletal muscle, multinucleated giant cells and collagen fibres.[13] The presence of multinucleated giant cells, re-generating and degenerating muscle fibres in addition to the typical clinical presentation is essential in making diagnosis.[13] Most cases of fibromatosis colli resolve spontaneously or in response to conservative management with surgical intervention only carried out when there is no resolution

after 1 year or in those with craniofacial abnormalities.[14] Many studies have shown that 80% to 90% of patients respond to conservative management with physiotherapy.[15] The conservative management consist of physiotherapy with heat, massage, stretching exercises for involved muscle, strengthening exercises for contralateral muscle, and occasionally the use of neck braces during sleep.[14] Do reported that when physiotherapy was commenced within the first 4 months, resolution occurred in approximately 3 to 4 months.[16] Passive stretching exercise is successful in more than 90% of cases when commenced within the first 3 months of life.[12] The index patient was commenced on stretching exercise at six weeks of age and the tumour was barely noticed after 3 months. Surgery is only required in about 5% of patients that presented early, and in up to half of those seen after 6 months of age.[12]

Conclusion

Fibromatosis Colli is a rare tumour of infancy. The aetiology of the disease is unknown, however, it can subterfuge a wide range of diseases. Its definitive diagnosis depends on the patient characteristic clinical presentations, non-invasive radiological investigations and fine-needle aspiration cytology. Thus, avoiding unnecessary surgery in a lesion which is commonly managed conservatively.

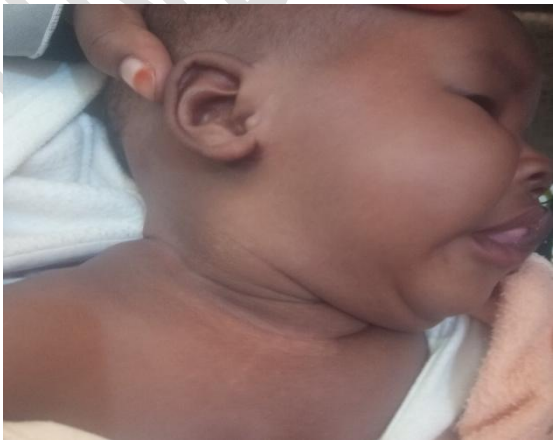

Figure 1: Picture shows tumour in the right sternocleidomastoid muscle

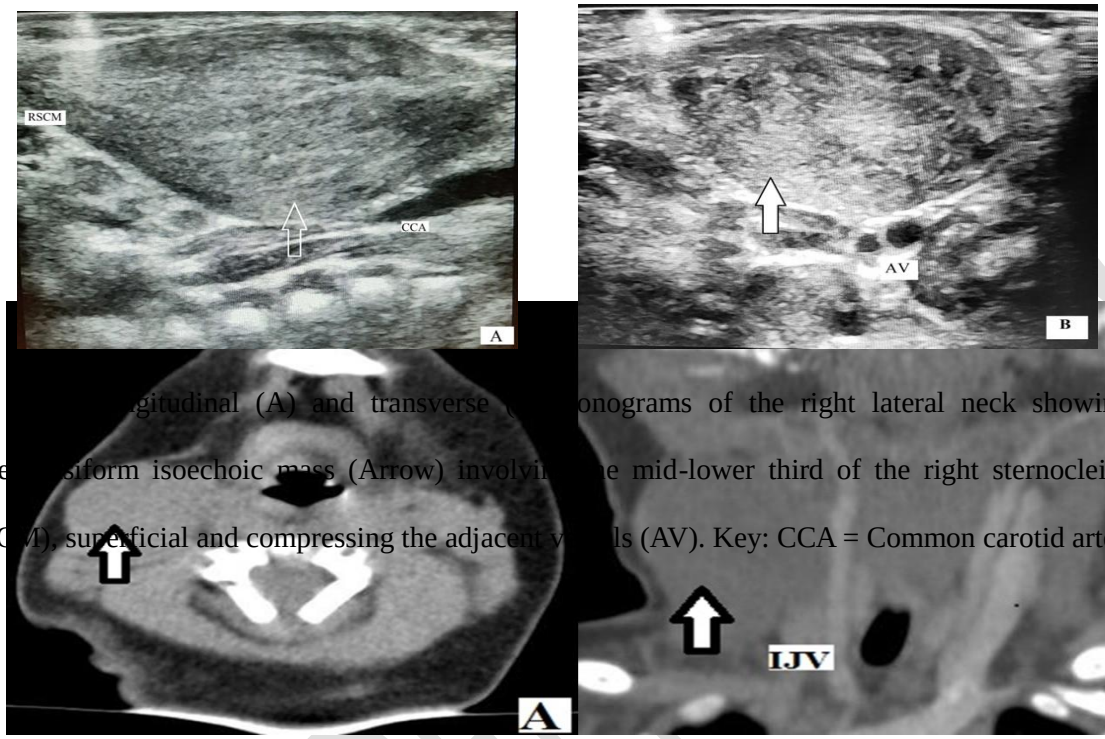

Figure 2(A [illegible] gitudinal (A) and transverse ([illegible] onograms of the right lateral neck showing a well circumscribe[illegible]siform isoechoic mass (Arrow) involvi[illegible]e mid-lower third of the right sternocleidomastoid muscle (RSC[illegible]M), superficial and compressing the adjacent v[illegible]ls (AV). Key: CCA = Common carotid artery

Figure 3(A & B): Axial pre-contrast (A) and coronal reformatted post-contrast(B) computed tomographic images showing a well-defined fusiform isodense mass (Arrow) in the mid-lower third of the right sternocleidomastoid muscle and compresses the adjacent internal jugular vein

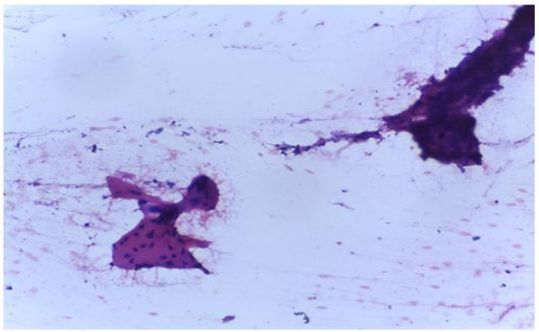

Figure 1: Smears show clusters of spindle shaped cells along with multinucleated giant muscle cell in a myxoid background, (Haematoxylin and Eosin stain X 200).

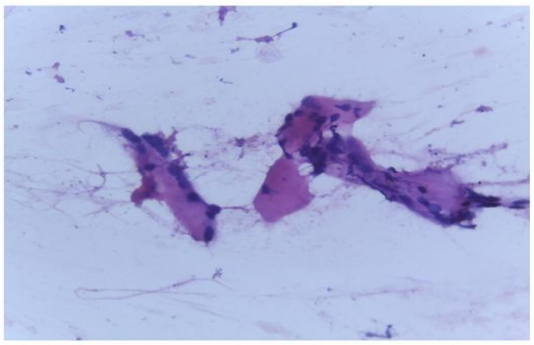

Figure 2: Smears shows muscle cell with a wispy cytoplasm, (Haematoxylin and Eosin stain X 200).

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