

Clinical and Diagnostic Approach to a Pediatric Patient Presented with a Lateral Neck Mass

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Abstract: Sternocleidomastoid muscle tumor of infancy, known as Fibromatosis colli is a relatively rare benign lesion that appears mainly unilaterally in the neck of an infant, with an incidence of 0.4% of births. It is one of the most common causes of congenital torticollis and more common in males. It appears in a newborn between the second and eighth weeks of life. It is usually unilateral (in the right sternocleidomastoid muscle) and occurrences on both sides of the neck are very rare. Over 95% of these tumors resolve spontaneously or with stretching exercises after 4-8 months of life. Diagnosis is based on patient history, clinical presentation and use of ultrasound. Invasive diagnostic methods such as a biopsy are not routinely used are reserved only in cases where there are diagnostic difficulties. Most cases have a good prognosis if therapy is started as early as possible and performed for as long as necessary. We report the case of a two-months-old boy, presenting a right lateral cervical mass. Clinical presentation, objective examination and ultrasound appearance strongly supported the diagnosis of Fibromatosis Colli. Conservative physical therapy with passive stretching exercises, massage and follow-up in dynamics was recommended.

Keywords: Fibromatosis Colli, Neck Mass, Congenital, Sternocleidomastoid, Pediatric, Infants

1. Introduction

Fibromatosis Colli is a benign sternocleidomastoid (SKM) muscle tumor that appears as a mass in the neck of an infant mainly in 2-8 weeks of life and start regressing at 4-8 months to fully resolve in the 1-2 years of life. [1] It is one of the most common causes of congenital muscular torticollis. WHO in 2020 has classified fibromatosis colli in the group of benign fibroblastic and myofibroblastic tumors, the cause of congenital muscular torticollis [2] These tumors are more common in male infants and are more commonly localized on the right side of the neck muscle. [3] The identification of a mass in the SKM muscle in the neonatal age requires a regional neck ultrasound to establish the diagnosis of Fibromatosis colli or to exclude it. Ultrasound remains the first and chosen examination, due to its low cost, non-invasive procedure, no need for sedation and has 100% sensitivity in making the diagnosis. [4]. Conservative therapy with physiotherapy after one month gives improvement in about 95% of infants. Children who are diagnosed late (after

12 months of age) and infants who do not show improvements (after 1 year) from physiotherapy surgical treatment is recommended. [1, 12]

2. Clinical Case

A two-months-old boy was hospitalized to the PICU of the “Mother Teresa” Hospital, University Center, Tirana, Albania with high fever, bronchiolitis and acute respiratory failure. Occasionally during objective examination the pediatrician noticed a tumefaction on the right side of the neck and presence of torticollis. The boy was born by vaginal delivery at 39 weeks of gestation, birth weight of 3100gr, height 50 cm and head circumference 35 cm. Cephalic birth position and mother does not refer use of forceps during birth. He is the first child of the couple, from nonconsanguineous parents. The mother denied having any infectious diseases during the pregnancy. Family history was negative for fibrous tissue tumors or congenital torticollis. Examination

showed non-tender neck swelling on the right side of the neck in the lower third of the sternocleidomastoid muscle, firm, non-tender, with reduced neck movement on the affected side. Clinical examination was otherwise unremarkable with no enlarged lymph nodes palpable elsewhere. Mother refers to that she noticed the mass in the baby's neck during the sixth week of life that has been increasing in size. A neck ultrasound was performed. On neck ultrasound was described a formation of the right sternocleidomastoid muscle with dimensions 38×16 mm with the presence of vascularization in its middle portion with heterogeneous echogenicity suggestive for fibromatosis of the sternocleidomastoid muscle or Fibromatosis Colli.



Figure 1. Right firm neck mass view, lateral cervical swelling without inflammatory signs.

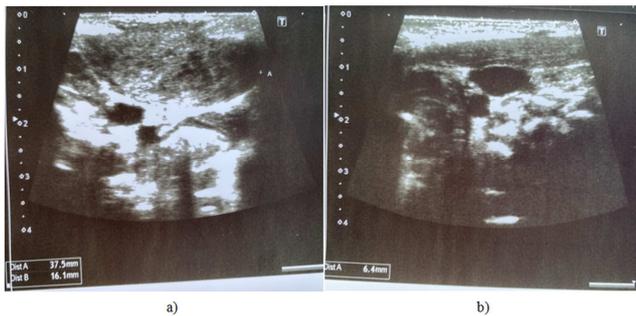


Figure 2. a, b Ultrasound images of the right neck of 2-months old boy with fibromatosis colli. Longitudinal images revealed a fusiform dilatation of SCM muscle (3.75×1.61 cm). The echostructure of the mass compared with the normal muscle.

The child was diagnosed with fibromatosis colli and congenital muscular torticollis, on discharge from the hospital was recommended conservative physical therapy with passive stretching exercises, massage and follow-up in dynamics and re-examination by the pediatrician.

3. Discussion

After encountering in our clinic the case of a 2-month-old boy with the presence of a cervical mass that was diagnosed with fibromatosis colli we review the published literature for diagnostic criteria, diagnostic methods and contemporary treatment methods recommended about fibromatosis colli. Fibromatosis colli was first described in the German literature in 1812 as a “sternocleidomastoid tumor” cause of congenital torticollis. [5] The presence of lateral cervical mass in an infant's neck can have different pathological,

acquired and congenital causes. Benign lesions should be distinguished from masses or lesions that hide serious problems in a child, and this should be well known by the pediatrician. Fibromatosis Colli is known as a benign SKM muscle tumor encountered in an infant. It is important that the SKM muscle tumor in an infant be differentiated from several pathologies such as: thyroid cyst, brachial cyst, cystic hygroma or cancerous lesions such as neuroblastoma or lymphoma. Fibromatosis colli is a common cause of neonatal torticollis as 20% of cases are related to torticollis, and in our case the patient also presents torticollis as in the literature. [6] It has a characteristic clinical presentation, it is observed in the neonatal period, usually in 2-8 weeks after birth but may be present at birth. In our case it was diagnosed in the eighth week of life, but the mother reports that she noticed it in the sixth week after birth and the mass has been gradually increasing. It is usually unilateral and occurrences on both sides of the neck are very rare, but in the literature there are some cases reported with bilateral forms of fibromatosis colli. [7] In a study it was found that right localization is encountered in 75% of cases and left in 22% of cases. [8] It is a benign lesion that is encountered in infancy, with an incidence of 0.4% of births. [9] 95% of cases have a good prognosis if therapy is started as early as possible and performed for as long as necessary. [10-12] The etiology remains unclear, but it is thought that there are some factors influencing the onset of the lesion. It has been observed that infants presenting fibromatosis colli had a history of traumatic birth, difficult birth, intrauterine misalignment, podalic presentation. [13]

Diagnosis is based on clinical presentation and history, use of ultrasound and invasive diagnostic methods such as a biopsy (which is not routinely used, but only in cases where there are diagnostic difficulties). The typical localization at the SKM muscle, usually localized in the lower 1/3, the characteristics during the objective examination and the age of the infant during the presentation helps us to define the diagnosis. Rapid regression of the mass in the first months of life with or without physiotherapy provides us for the diagnosis of fibromatosis colli by avoiding unnecessary further examinations. For the follow up, neck ultrasound remains the chosen modality. Ultrasound features of the lesion of fibromatosis colli are diffuse or focal fusiform enlargements of the SKM muscle, localized mainly in the lower 1/3, with defined borders and homo or heterogeneous structure. [11, 14, 15] Fine needle aspiration biopsy is a safe method that can be used to confirm the diagnosis in cases where the neck mass in an infant is suspicious and other examinations besides ultrasound are needed. [16] While excisional biopsy is better to avoided and should be reserved only for those cases where we have a high clinicopathologic suspicion of malignant tumors. Regarding the histopathological examination of Fibromatosis Colli, cytologic features observed are bland-appearing fibroblasts and degenerative atrophic skeletal muscle changes. [17] High-resolution imaging such as CT and MRI may be helpful, but are not routinely used for diagnosis. MRI helps

to accurately determine the extent of the lesion and the relationship with the SKM muscle. [11] Fibromatosis Colli are lesions that increase in size in the first month of life until they reach a maximum size and then the mass gradually decrease. 95% of them resolve spontaneously or with conservative therapy after 4-8 months of life, to disappear completely in the 1-2 years of life [1, 12]. Conservative therapy consists in: Passive stretching exercises, repositioning procedures and massage. The success of conservative therapy depends primarily on the age of the patient at the time of diagnosis and the earliest possible start of physiotherapy. In some cases where the tumor persists despite physiotherapy and in those cases where we have deformities of the face or body of children, surgical correction is recommended. [18-20].

4. Conclusion

In conclusion, we can say that in the presence of torticollis and a mass in the neck of an infant a few weeks after birth or since birth the pediatrician should consider Fibromatosis Colli. A careful clinical history, clinical findings during objective examination and the use of neck ultrasound data can determine the diagnosis of Fibromatosis Colli avoids the use of unnecessary invasive procedures such Fine needle aspiration biopsy or surgery and use of MRI. Ultrasound remains chosen examination for diagnosis and follow-up. The evolution of fibromatosis colli is spontaneous regression or under physiotherapy for a few months.

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