Case Report / Olgu Sunumu

Fibromatosis Colli: A rare neck mass in infancy

Fibromatozis colli: Bebek çağında nadir bir boyun kitlesi

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Abstract

Fibromatosis colli (FC), also known as sternocleidomastoid (SCM) tumor of infancy, pseudotumor of infancy, and congenital muscular torticollis, is categorized as a benign fibroblastic proliferation in the 2002 World Health Organization (WHO) classification of soft-tissue tumors. It is the most frequent cause of congenital torticollis. Its etiology is unknown and it is likely related to birth trauma or results from an in utero fetal head position [1-3]. Ultrasonography (US) is the imaging method of choice for evaluating congenital neck lesions in children, and the results can often suggest the correct diagnosis. Computed tomography (CT) or magnetic resonance imaging (MRI) is sometimes required to define the extent of the lesion [4, 5]. In this article, we describe the US pattern of this entity in order to facilitate its recognition and to avoid more invasive methods for diagnosis.

Keywords: Fibromatosis colli, sternocleidomastoid muscle, ultrasonography

Özet

Fibromatozis kolli bebeklere gören fibromatozisin sternokleidomastoid kasta gören nadir bir formudur, yaşamın ilk 1-8 haftasında gören, ağrı olmayan ve tortikolise neden olan bir boyun kitlesidir. Ultrasonografi ile fibromatozis kolli tanı konulan, sağ boyun kitlesi ve tortikolis ile başvuran 6 haftalık bir kız bebek olgusu sunulmaktadır.

Anahtar sözcükler: Fibromatozis kolli, sternokleidomastoid kas, ultrasonografi

Introduction

Fibromatosis colli (FC), also known as sternocleidomastoid (SCM) tumor of infancy, pseudotumor of infancy, and congenital muscular torticollis, is categorized as a benign fibroblastic proliferation in the 2002 World Health Organization (WHO) classification of soft-tissue tumors. It is the most frequent cause of congenital torticollis. Its etiology is unknown and it is likely related to birth trauma or results from an in utero fetal head position [1-3]. Ultrasonography (US) is the imaging method of choice for evaluating congenital neck lesions in children, and the results can often suggest the correct diagnosis. Computed tomography (CT) or magnetic resonance imaging (MRI) is sometimes required to define the extent of the lesion [4, 5]. In this article, we describe the US pattern of this entity in order to facilitate its recognition and to avoid more invasive methods for diagnosis.

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Case

A 6-week-old female baby presented with right neck mass and torticollis. The mass was firm and well circumscribed, and found within the right SCM muscle. She also had torticollis with her chin and face turned to the left shoulder. The remainder of her physical examination was normal. Laboratory results were normal limits. Ultrasonographic examination of the neck demonstrated a homogenously diffuse enlargement of the right SCM muscle without focal mass lesion. The lesion was slightly hyperechoic according to the muscle (Figures 1a 1b). There was no mass effect and no abnormal cervical lymphadenopathy. Cervical vascular structures were found to be normal. Clinical and imaging findings were typical of FC. No treatment was undertaken, and the lesion showed gradual resolution in the ensuing months.

Discussion

FC is a rare form of infantile fibromatosis occurring in the SCM muscle. It usually occurs in the 1st to 8th weeks of life with a history of a non-tender neck mass and torticollis (tilting of the head toward the same side with rotation of the chin toward the opposite side) in 14 to 20% due to muscle contraction. It is a firm, painless, discrete, fusiform mass within the
SCM typically measuring between 1 and 3 cm. in diameter [1, 3, 4, 6]. FC affects male patients slightly more often than female patients. The mass is typically located in the middle or inferior portions of the SCM muscle. FC is usually unilateral and generally found on the right side. Our patient had the lesion on the right side of the neck. Bilateral involvement is rare. In addition to torticollis, patients may present with lytic clavicular lesions, ipsilateral mandibular asymmetry, plagiocephaly, occipital condyle asymmetry, elevation of the ipsilateral clavicle and shoulder, facial deformities and postural cervicothoracic scoliosis [1-4].

The etiology of FC is unknown. One possibility may be intrauterine malposition causing restriction of blood supply to the sternal and middle fibers of the SCM. Another possible cause is birth trauma, particularly through breech deliveries, with pressure necrosis and organizing fibrosis. Compartment syndrome causes necrosis of the SCM muscle and resultant fibrosis [4, 6-8].

US is the principal diagnostic tool for FC and has 100% sensitivity. The infant neck is best imaged with a high-frequency (7-MHz) transducer. A linear array transducer is preferred in order to image a larger near field of view, since most structures of interest are relatively superficial. Ultrasonographic findings include a uniformly isoechoic or slightly hypoechoic mass within the muscle or a homogeneously enlarged SCM muscle without a focal mass. The mass can be hypo-, iso-, or hyperechoic, depending on age. With real-time ultrasonography, the lesion can be shown to move synchronously with the SCM muscle. CT or MRI should not be necessary when both the ultrasonographic and the clinical findings are compatible with FC, as in our case [3, 5-7].

In addition to FC, the differential diagnosis of soft-tissue masses in the neck of a neonate includes neuroblastoma, rhabdomyosarcoma and other sarcomas, lymphoma, cystic hygroma, and branchial cleft cyst. Lesions that are more often midline or near the midline include dermoid cyst, teratoma, or thyroglossal duct cyst. The most useful imaging findings that suggest that the lesion is not FC is extension of the mass beyond the SCM muscle. Additionally, neuroblastoma and rhabdomyosarcoma characteristically have an infiltrative appearance, with irregular margins. They may also contain calcifications. Lymphoma is suggested by the presence of oval masses corresponding to enlarged lymph nodes along the cervical lymph node chain [1, 4, 5-7, 8, 9].

Biopsy or surgical intervention should not be necessary if the imaging findings and clinical setting are appropriate for the diagnosis of FC. Treatment primarily consists of physical therapy. There is usually gradual improvement after the institution of physical therapy, with increased range of motion over a period of 4 to 6 months [1-3, 6, 7].

In conclusion, the diagnosis of FC can be made by only US. In most cases, MRI, CT, fine needle aspiration, and histologic examination are not necessary.

References