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STERNOCLEIDOMASTOID PSEUDOTUMOR OF INFANCY: A REPORT OF THIRTEEN CASES

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Sternocleidomastoid tumor of infancy (SCMTI) is a rare cause of benign neck masses in neonates and infants. It has to be differentiated from other congenital space-occupying lesions in the cervical region.
Patients and methods: **The files of 13 infants with a mean age of 6 weeks, presenting with a lateral neck mass at Sahloul University Hospital in Sousse (Tunisia) between 2007 and 2009 were retrospectively studied. All of them underwent physical and ultrasonographic examination. MRI was performed in only one case.**
Results: **Ultrasonography (US) showed a soft tissue mass of sternocleidomastoid muscle (SCMM), or a homogenously enlarged muscle without any focal mass. MRI revealed a fusiform enlarged muscle. Diagnosis of SCMTI was established in all cases. Conservative treatment was recommended in all cases with physiotherapy in 2 cases.**
Conclusion: **US is the best imaging modality for the diagnosis of SCMTI and the first one to be performed. Additional diagnostic imaging modalities are unnecessary in most of the cases.**

Key-words: Infants – Head and neck neoplasms, US.

Diagnosis and management of neck masses in infants and children are frequently challenging for radiologists, paediatricians and otolaryngologists. Only a thorough knowledge of the different clinical entities that can cause neck masses in the paediatric population can insure immediate and appropriate treatment. Sternocleidomastoid tumor of infancy (SCMTI), also known as fibromatosis colli, is the most common cause of neck masses in the neonatal period (1). It is a thickening of the sternocleidomastoid muscle (SCMM). It presents mostly between the second and eighth week of life when the parents notice a firm tumor in the neck, which can be followed by a characteristic torticollis (2).

US is the best imaging modality for diagnosis. MRI or CT are sometimes performed when diagnosis is uncertain or when this entity is unknown by the clinicians (2).

We report 13 cases of SCMTI explored by US, and MRI in one case, in order to facilitate the recognition of this entity.

Patients and methods

Thirteen cases of SCMTI were diagnosed in our institution during a-three-year period between 2007 and 2009, including 10 boys and 3 girls aged between 2 and 11 weeks with a mean age of 6 weeks when the diagnosis was established. In 9 cases, the mass was discovered by the parents and in the 4 remaining cases by the pediatrician at the time of the systematic consultation at the 40th day of life.

All patients had physical examination, and their families were asked about birth conditions. In 12 cases, it was a low-way delivery, and one patient had been delivered by caesarean section because of breech presentation. Premature birth was noted in 2 cases, but there were no forceps deliveries or difficult labor in all cases. Birth weight ranged between 2,600 and 4,250 g.

All patients presented with a neck mass which appeared in the first weeks of life. A torticollis was associated to in 2 cases.

US was performed for all of them, and MRI in only 1 case because of uncertain diagnosis.

Results

The mass was firm and painless. It was located in the left side in 9 cases and in the right side in 4 cases. Torticollis was noted in 2 cases.

Neck US showed hypoechogenic or isoechogenic, homogeneous and fusiform mass of the SCMM in 8 cases, and a heterogeneous mass in 2 cases (Fig. 1). In 3 cases, the SCCM was homogeneously enlarged without any focal mass. In these cases, the fibrillar structure of the muscles was maintained (Fig. 2).

The tumor was located in the caudal part of the muscle in 10 patients, and in the middle part in the remaining 3 cases. The size of the lesion varied between 2 and 4 cm in great diameter, with a mean of 2,7 cm.

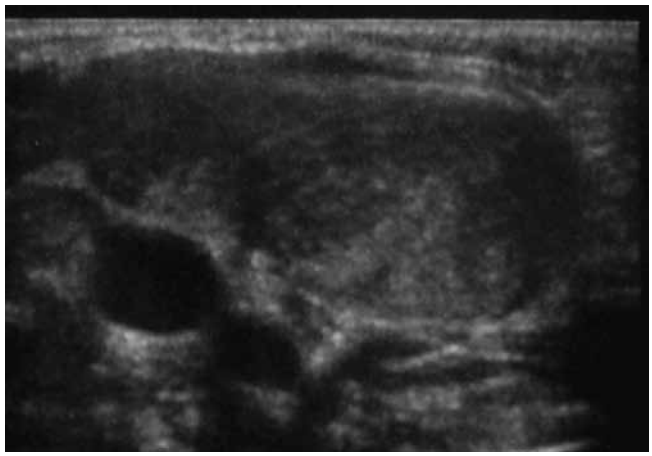

Fig. 1. — Sternocleidomastoid tumor of infancy in an-8-week old girl. Axial sonography shows faintly heterogeneous mass within the left SCCM.

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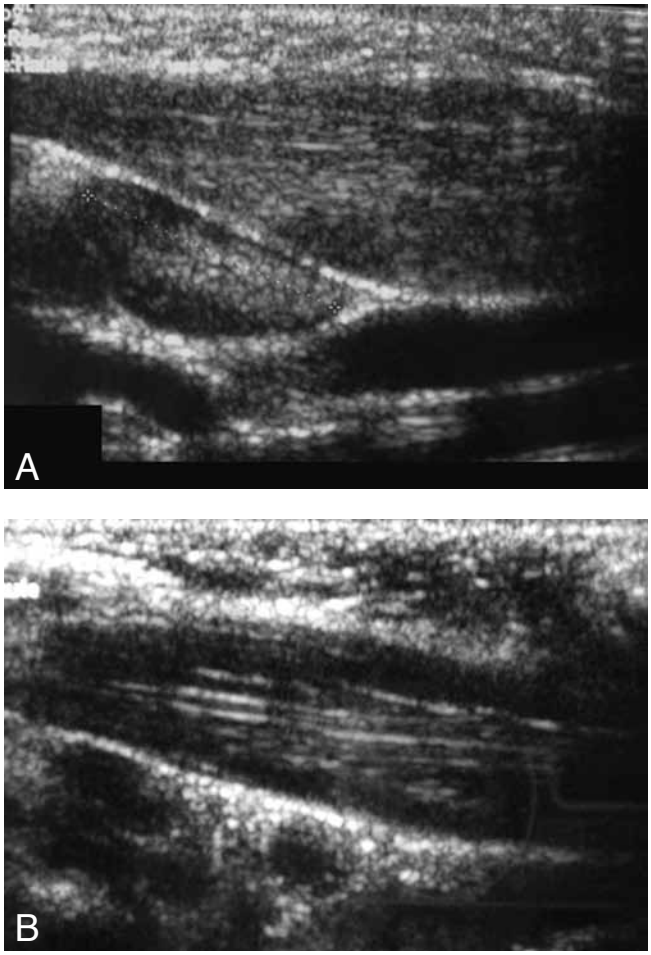


Fig. 2. — Sternocleidomastoid tumor of infancy in a-6-week old boy. Sonography shows in (A) homogeneous enlargement of the right SCMM with lymph nodes in the posterior cervical space. In (B) decrease of abnormalities 40 days later.

All tumors were well delineated from the surrounding structures. Cervical vascular structures were normal. In 2 patients, physical examination revealed cervical lymph nodes and one of them required MRI which was performed 15 days later, and showed an enlargement of the right SCMM without focal mass or abnormal enhancement. Lymph nodes disappeared (Fig. 3).

Diagnosis of SCMTI was made in all cases. Physiotherapy was recommended as the initial treatment in 2 patients who presented with torticollis.

Six patients were followed clinically, and 3 by US during six months. Nine patients had adequate follow up which revealed complete regression of the neck mass and torticollis. The four remaining patients were not followed, and we presume that the mass resolved spontaneously.

Discussion

SCMTI is a rare form of infantile fibromatosis that occurs solely in the SCMM with a reported incidence of 0.4% live births (1, 3). The disease is usually unilateral, slightly more common on the right side and affects both sternal and clavicular heads of the muscle. Bilateral involvement is extremely rare (2). SCMTI affects male patients slightly more often than females (4, 5).

Most cases show no abnormality at birth, but the SCMTI appears between the second and eighth week of life as a firm, painless, fusiform soft-tissue mass in the lower third to the second third of the SCMM, typically measuring between 1 and 3 cm in diameter (2, 6). The mass may increase in size for several weeks, then stabilizes in size for a few months, and finally decreases spontaneously by 4 to 8 months of age (6). Torticollis is seen in 14% to 20% of patients due to contraction of the SCMM. It has a characteristic appearance: The head is rotated and tilted toward the side of the mass, and the chin is turned away from the affected side. This is secondary to fibrosis, shortening of the affected muscle, and its inability to keep space with the growth of the normal SCMM (1, 2).

Although the exact cause is unclear, it is likely related to birth trauma, with greater than 90% of cases associated with a difficult or forceps delivery (1, 4). One theory contends that traumatic compression of the neck during delivery could result in pressure necrosis or occlusion of the venous outflow system with subsequent development of muscle edema, degeneration of fibers, and fibrosis.

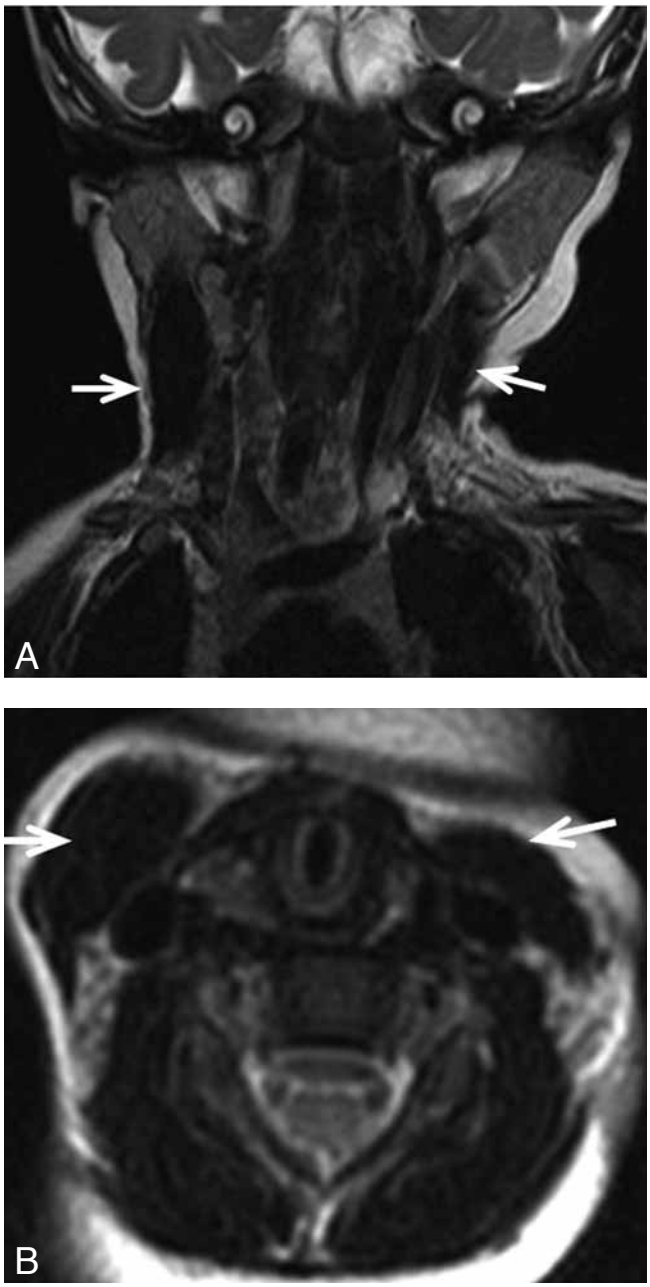
Davids and co-workers (7) have suggested that the lesions result from an in utero fetal head position, which causes selective injury to the SCMM. Such injury leads to the development of a secondary compartment syndrome resulting in pressure necrosis and fibrosis within the muscle.

A genetic component may also play a role, as 11% of patients had a positive family history (6, 8). In our series, there was not any apparent cause of this disorder in all patients. Ten of them come from the same village, but they were born in four different centers. We believe that a genetic factor might be considered.

In most cases, the clinician can make the diagnosis of SCMTI by careful history and physical examination. A detailed obstetrical history should be obtained as there is a well recognized association between SCMTI and primiparous births, breech presentations, forceps deliveries, and difficult labor.

Imaging findings in this disorder must be differentiated from other causes of neck masses in this age group. When the abnormality is not recognized, inappropriate and invasive procedures may be performed such as needle biopsy, open biopsy, and even resection of the SCMM. Only recognition of this entity can prevent unnecessary diagnostic and therapeutic procedures (1).

US is the best imaging modality for diagnosis and should be the first examination to be performed, due to the proximity of the lesion to the skin and its lack of radiation (1, 2). The lesions measure several centimetres in diameter (9). Various US findings



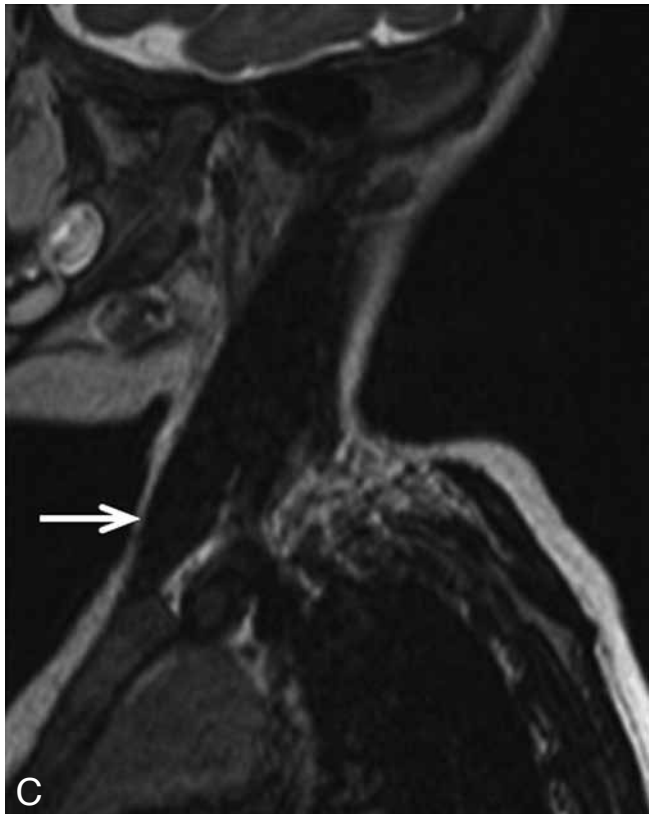


Fig. 3. — The same case as Fig. 2. MRI: Coronal (A) and axial (B)T2 weighted images. Homogeneous enlargement of the right SCMM, the left one is normal (arrows). Sagittal T2 weighted images (C) show involvement of the sternal side of the muscle (arrow).

have been described, ranging from a homogeneously enlarged SCMM without a focal mass to a hypoechogenic mass with ill-defined or well-defined margins in the substance of the SCMM (1, 9).Typically a non homogeneous, echopoor tumor with increased perfusion can be shown (10). With real time sonography, the lesion can be shown to move synchronously with the SCMM (2, 4).

MRI or CT are not necessary when both US and clinical findings are compatible with SCMTI. MRI and CT reveal similar features, including fusiform thickening, well-defined margins, and mild mass effect on adjacent structures. The involved portion of the muscle is better defined on MRI or CT than sonography (10).

The tumors are commonly located in the distal 2/3 of the SCMM and show good delineation from surrounding structures (10).

MRI may show signal intensity characteristics. On T2-weighted MR images, the enlarged muscle demonstrates diffuse abnormal high signal intensity greater than that of fat, and no focal discrete mass may be noted. Decreased signal intensity of the mass in T2 weighted images can be seen because of presence of fibrous tissue (2). The MR Imaging appearance has been described by Davids and co-workers (7). All cases in their series demonstrated mild enlargement of the lower third of the SCMM with diffuse abnormal high signal intensity, greater than that of fat on T2-weighted images. None of the cases in their series demonstrated a focal discrete mass.

MRI also helps demonstrate lack of involvement of surrounding structures and absence of lymphadenopathy, airway compression, vascular encasement, bone involvement, intracranial/intraspinal extension associated with other neck masses or other features that would suggest a more aggressive process (2, 10). The multiple lymph nodes revealed by US in one of our cases, were not found in MRI performed 2 weeks later. We think that these lymph nodes were associated to another banal inflammatory disorder.

Although radiographs are usually normal, lytic lesions within the head of the clavicle at the attachment of the SCMM have rarely been reported (1).

If imaging studies show other findings such as inhomogeneous

muscle density or echo pattern, a lesion within the SCMM with irregular margins, a mass extending beyond the margins of this muscle, or significant regional lymphadenopathy, then other alternative diagnoses should be considered.

In addition to SCMTI, the differential diagnosis of soft-tissue masses in the neck of a neonate includes sarcoma, lymphoma, cystic hygroma, and branchial cleft cyst. Lesions that are often midline or near the midline include dermoid cyst, teratoma, or thyroglossal duct cyst. However, rapid growth of the tumor and increased thickening of SCMM should raise the possibility of another tumor, such as rhabdomyosarcoma, or a tumor originating from outside the muscle but infiltrating the surrounding structures, such as neuroblastoma (2, 6).

Kumar and co-workers (11) think that non invasive diagnosis of SCMTI can be made by fine needle aspiration cytology alone; invasive diagnostic and therapeutic measures are best avoided. Excision biopsy may not be necessary and should be reserved for cases with a strong clinicopathologic suspicion of malignancy.

Although pathologic specimens are rarely encountered today, typical changes observed at histologic evaluation include alternating skeletal muscle fibers that have undergone atrophy or degeneration and evolving scar like fibroblastic-myofibroblastic proliferation (3, 8).

SCMTI usually resolves with conservative management in most of the cases, including clinical observation after the diagnosis is established on a clinical, radiologic, or less frequently histologic basis (10).

In more than 70% to 90% of cases, lesions regress spontaneously, and secondary contractures resolve within 4 to 8 months (4). Physiotherapy is recommended as the initial treatment. Surgical intervention is required in 10% to 15% of cases, and is indicated in patients with severe refractory disease after 1 year of age to prevent permanent contracture and plagiocephaly (2, 10).

Surgical treatment consists of proximal or distal release of the SCMM.

Sixteen to seventy percent of patients with persistent torticollis after resolution of the SCMTI will develop cranial or facial asymmetry if the torticollis is not corrected (2, 4, 10).

Conclusion

SCMTI is a well recognized clinical entity that typically responds to conservative therapy. Diagnosis is often made clinically with a detailed history and physical exam. US is the best imaging modality for diagnosis. Variations in its appearance should be kept in mind and should not prevent the correct diagnosis as long as the abnormality is intramuscular and adjacent soft tissues are normal. MR Imaging and CT are indicated in unusual cases when diagnosis is uncertain, and to evaluate masses that do not respond to therapy. Fine needle aspiration and histologic examination are not necessary and should be reserved for lesions with clinical signs and imaging findings inconsistent with SCMTI.

The recognition of this entity can prevent unnecessary diagnostic and therapeutic invasive maneuvers.

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