Torticollis in children: A pictographic review


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Abstract. Torticollis describes the clinical finding of an abnormal positioning of the head related with the body axis, with cervical rotation and contralateral tilt of the head, which is usually secondary to an involuntary contraction of the cervical musculature been sternocleidomastoideus muscle (ECM) the most important component. As a clinical sign, differential diagnosis is very broad, and may be secondary to multiple causes. Pediatric population has a particular spectrum of diseases which differs from adults. The aim of this pictorial review is to evaluate some torticollis causes in the pediatric setting recording main imaging findings and their contribution to the clinical diagnosis.
Keywords: Children, Imaging, Torticollis.

Resumen. La tortícolis describe el hallazgo clínico de una posición anómala de la cabeza respecto del eje corporal, con rotación cervical e inclinación contralateral de la cabeza, que habitualmente es secundaria a una contracción involuntaria de la musculatura cervical con compromiso predominante del músculo esternocleidomastoide (ECM). Como signo clínico su diagnóstico diferencial es muy amplio, pudiendo ser secundario a múltiples causas. En la edad pediátrica el espectro es particular y difiere de la forma reconocida en adultos. El objetivo de esta revisión pictográfica es evaluar algunas de las causas de torticolis en la edad pediátrica y analizar los principales hallazgos imaginológicos y su aporte al diagnóstico clínico.
Palabras clave: Imágenes, Niños, Torticolis.

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Material and methods
A retrospective review of patients under 15 years of age who had torticollis amongst their symptoms on admittance, between june 2007 and july 2011, evaluated with simple radiography, ultrasound, CT scan or magnetic resonance imaging (MRI).

Results
The causes of torticollis may be situated in bone structures at the base of the skull and the cervical spine, in the soft areas of the neck and the CNS. In the time interval examined, patients presented with secondary torticollis to pathologies not only in soft tissue but also in bone structures, as summarized in Table I. Below are images and comments of selected cases for each disease, highlighting the imaginological characteristics in the different kinds of study.

A) Soft tissue of the neck
Fibromatosis Colli
Congenital muscular torticollis (CMT) or fibromatosis Colli is a pathological condition characterised by morphological and functional changes of the sternocleidomastoid muscle (SCM), whose etiopathogeny has not yet been specified. It clinically manifests around the third week of life as a cervical mass, which can be tender or by a tilt of the head toward the affected side. It is more common in association with traumatic delivery in relation to the use of forceps, in breach presentation and primiparas. Treatment is based primarily on physical therapy and its clinical evolution is generally self-limiting, but may leave permanent damage if left untreated.

Ultrasonography is useful in the diagnosis of congenital muscular torticollis. Findings consist of a
relatively homogeneous increase in volume of the affected muscle, with an appearance of a well demarcated solid mass, which usually compromises the upper or middle thirds of the muscle. Its echogenicity tends to be slightly larger or smaller than the adjacent healthy muscle and may present increased vascularization during color Doppler scanning (Figure 1).

**Sternocleidomastoid muscle hematoma**

Congenital torticollis, described previously, must be distinguished from other less frequent infiltrative conditions of the SCM, such as fibrosis and hematomas. Most SCM hematomas occur in the neonatal period, and are secondary to obstetric trauma, where the stretching of the muscle generates a bleeding inside the muscle sheath, which is usually resolved spontaneously. The later presentation usually occurs in the context of traumas of moderate to high energy (Figure 2).

![Figure 1. 20 day old patient, with a marked tilt of the head to the right. Ultrasound, longitudinal image of the neck, demonstrates increased volume (a) and vascularization (b) of the left SCM with respect to the contralateral (c).](image1)

![Figure 2. 3 year old patient, pain and increased right cervical volume after a high-energy direct contusion (bicycle accident). Ultrasound shows definition loss of the muscle planes and an increase of the right echogenicity in comparative cross-sectional image (a) and longitudinal (b). CT shows an increase in the muscle density (c) and of the surrounding soft tissue (d).](image2)

**Sternocleidomastoid muscle fibrosis**

Corresponds to an uncommon pathology with a poorly defined etiology; the term is often used as a synonym for congenital torticollis, but not only their clinical presentation but also imaginologically they differ. It presents beyond the neonatal period, in patients with persistent head lateralization and often with a history of multiple kinesic treatments with little or no improvement (Figure 3).
Cervical adenitis

The term “cervical adenitis” has been used as a synonym for the presence of adenopathies. Strictly speaking this should be reserved for those conditions which are accompanied by inflammatory changes of the adjacent tissue, to distinguish it from reactive lymphoid hyperplasia which frequently accompanies orofacial infections in children. Acute cervical adenitis is mainly related to bacterial infections. They are normally adenopathies of 2-3 centimetres, painful on palpation and unilateral, in a usually severe condition for 5 days or less. Over 80% of the cases are due to Staphylococcus aureus and Streptococcus pyogenes. Infections by these bacteria are more common in preschool children, secondary to an oropharyngeal or cutaneous outbreak. The most frequent complication is abscess formation, appearing in 10 to 25% of the cases. Anaerobic infections usually occur in older children with dental pathology (Figure 4). Jugular thrombosis, as part of the septic process or Lemierre syndrome, is a rarer complication. It has been described more often related to infectious odontogenic or pharyngeal diseases (Figure 5).

Retropharyngeal abscess

This corresponds to the potentially lethal infection of the retropharyngeal space, most often by an infectious etiology, in general, secondary to the spreading of oropharyngeal infections to the lymphatic structures of the retrofarinx, also being secondary to trauma, iatrogenesis or foreign bodies. Up to 75% of the cases occur in children under 5 years of age due

Figure 3. 7 year old boy with a history of torticollis since birth. Ultrasound shows thinning and altered echostructure of the right SCM, thinned and echogenic, in comparative images in axial plane (a) and longitudinal (b).

Figure 4. 4 year old patient, no relevant morbid history, right cervical volume increase associated with fever, with cervical lateralization. Ultrasound shows extensive right submaxillary adenopathic conglomerates, with anechogenic collections without vascular flow and inflammatory changes of the adjacent adipose tissue (a). CT shows right submandibular adenopathy with a central necrosis area (b).

Figure 5. Lemierre syndrome. 18 month old patient with pharyngeal infection, cervical adenophlegmon and thrombosis of the jugular. Ultrasound shows adenopathy with a necrotic area (a), increased diameter of right internal jugular vein, with hypoechogenic endoluminal content and no flow on color Doppler image (b).
to the proportional increase of lymphatic tissue in this location at this age, which later regresses. The clinical presentation is variable, including dyspnea and stridor, drooling, meningeal symptoms or marked sepsis. The imaginological findings with simple radiography are limited, they may include an increase of the soft prevertebral tissue, being the rare presence of bubbles, the only pathognomonic sign of abscess. CT and MRI show increased volume of the retropharyngeal space, associated with the presence of a collection that shows annular impregnation with the use of contrast, with associated displacement of the airway and of the parapharyngeal spaces and carotids. Treatment is emergency surgical drainage, owing to the high risk of spreading to the mediastinum and its complication with mediastinitis, being often fatal (Figure 6).

**Thyroid abscess**

The thyroid abscess is a rare condition and a potentially fatal endocrine emergency. It represents 0.1 to 0.7% of thyroid pathologies. Thyroid infection can result from hematogenous or lymphatic spread, or from contiguity to an infection of the neck or oropharynx, by the presence of a foreign body or esophageal perforation. In children thyroid infections rarely originate in the gland itself, the most frequent cause is being the existence of anatomical alterations that allow the passage of infectious agents from the respiratory tract to the gland, the most common being the pyriform sinus fistula. The clinical condition is characterized by dysphagia, dysphonia, pain, fever and increased volume of the area; if the condition progresses, obstructive airway compromise and sepsis can exist. Thyroid function is normal in 83% of the cases, with isolated reports of hyperthyroidism. Ultrasound is the study of first choice for its low cost and invasiveness, showing a hypoechogenic parathyroid area, compatible with an abscess. CT and MRI are able to demonstrate thyroid and parathyroid commitment, and eventually the presence of a fistula, directly or using air as a contrast medium. CT is superior in demonstrating the presence of air in the fistula route and in defining the thyroid commitment, so this technique is preferred for assessment of the extent of the inflammatory process and its complications. The etiology is varied, the Gram-positives being the most frequent. In immunosuppressed patients, opportunistic agents should be suspected, such as Pneumocystis jirovecii and fungi. Once over the acute process, it is imperative to rule out the presence of a pyriform sinus fistula. This is a rare anomaly during the embryonic development of the 3rd and 4th pharyngeal pouches and whose exact origin is unknown. It extends from the apex of the pharynx to the parathyroid region,

**Figure 6.** 8 month old patient, upper respiratory infection and with small cervical adenopathies and cephalic lateralization to the right. Contrast-enhanced CT shows left retropharyngeal collection, with area of low density and peripheral enhancement in axial (a) and longitudinal (b). Cervical MRI confirms hypointense collection in T1 axial image (c) and hyperintense in sagittal T2-weighted (d) and marked peripheral reinforcement after the gadolinium injection (e), consistent with an abscess.
ending up in the thyroids or adjacent to this, which allows bacterial infection in or around the gland. Since it is a congenital anomaly, the development of a secondary thyroid abscess to a pyriform sinus fistula usually occurs in children. In over 90% of the cases it occurs on the left side. Treatment consists of broad-spectrum antibiotic therapy and surgical drainage in the acute phase, with a subsequent fistulectomy on a second occasion (Figure 7).

**B) Bony structures of the skull base and cervical column**

**Plagiocephaly**

This is a disorder characterized by an asymmetric distortion of the skull, by the unilateral or bilateral flattening of the occipital region. Among the forces that may determine the skull deformity are included various factors such as the position in utero, continuous support during sleep, poor psychomotor stimulation, for example, that conditions the persistent posterior support of the skull (positional plagiocephaly), keeping open the posterior sutures as opposed to the cases of synostotic plagioccephaly.

A high percentage of these patients suffer from congenital torticollis, and may also develop an addictive positioning of the head secondary to the cranial flattening. When there are doubts in the diagnosis, a simple radiograph of the skull should be performed to display the permeability of the parieto-occipital sutures, and thus rule out rare forms associated to craniostenosis. The preferred technique is cranial CT with 3D reconstruction, to allow to characterize and quantify the degree of plagiocephaly through the use of asymmetry indices, and also to allow to observe a slight enlargement of the subarachnoid space, an associated finding in up to 25% of the cases (Figure 8).

**Sprengel deformity**

The musculoskeletal abnormalities of the shoulder girdle can also be associated with cervical alignment alterations due to the dynamic imbalance between the forces, including in this group fundamentally those causes of asymmetric muscular commitment of the pectoral muscles and serratus anterior. Sprengels deformity or congenital undescended scapula corresponds to one of the causes of the clinical finding of “winged scapula”. This abnormality occurs due to failure of the normal descent thereof toward the chest wall between the 9th to 12th weeks, and is shown by a an evident lower pole of the scapula and a higher upper pole, which may even be level with the upper cervical vertebrae. All of these factors contribute to the significant limitation of the scapulothoracic mobility presented by these patients and may be associated with other skeletal disorders. The so called “bone omovertebral” is found in about one third of the cases of Sprengel’s deformity, and extends from the medial border of the scapula to the spinous processes of the cervical vertebrae C5 to C7 (Figures 9, 10).

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**Figure 7.** 8 year old patient, surgical history for cervical adenitis at 3 years. Currently odynophagia, anterior cervical pain and limited mobility, cephalic left tilt and adenopathies in both anterior jugular chains. Ultrasound shows poorly defined hypoechogenic area in upper third of left thyroid lobe (a) not vascularized (b) with suprathyroid extension through a pathway of 4mm (c). Contrasted CT confirms left thyroid collection (d) with pyriform sinus fistula in coronal reconstruction (e).
Figure 8. 7 month old patient, congenital torticollis without response to usual therapy. CT scout view displays alterations of the cephalic alignment with a tilt to the left (a), volumetric reconstruction, upper view shows slight right frontoparietal flattening with sutures of normal appearance (b).

Figure 9. 10 month old patient, cervical tilt to the right and functional limitation of arm and left shoulder. Simple radiography shows a smaller left scapula, elevated and rotated (a), best demonstrated in CT volumetric reconstruction (b).

Figure 10. 5 year old patient, carrier of Klippel-Feil syndrome with Sprengel deformity. Bone omovertebral in lateral cervical spine radiographs (a) and frontal (b) with fusion of posterior elements.
Langerhans cell histiocytosis

This is a rare disorder characterized by the proliferation and accumulation of histiocytes and eosinophils in various tissues, which comprises several distinct clinical entities, among which the most frequent corresponds to the so called eosinophilic granuloma (benign form that includes isolated monostotic bone involvement). This condition, with a not well defined etiopathogenesis, can occur at any age but presents a clear predominance in the pediatric age. In eosinophilic granuloma any bone can be compromised, but there is a predilection for flat bones, including amongst those the most frequently affected is the skull, not only the shell but also the base of the skull. Case symptoms will depend on the compromised bone, ranging from local pain and volume increase to neurological symptoms (e.g. when there is commitment of the petrosal process of the temporal bone). The findings are variable according to the stage of the disease, with aggressive looking osteolytic lesions in the acute phase (malignant periosteal reaction, soft tissue mass, etc.), which acquire a more benign aspect toward the intermediate and chronic phases of the disease (sclerotic border, unilamellar periosteal reaction, etc.). In the skull the lesions affect the external and internal tables with a characteristically varied involvement, and a lack of significant periosteal reaction. Other findings include the presence of bone infiltration, which should be distinguished primarily from the infection in children. CT is indicated to characterize bone involvement, and MRI to better define the possible extension to the adjacent soft intra- and extracranial regions (Figure 11).

Figure 11. 6 yearold patient, non-traumatic neck pain associated with torticollis and limitation of right rotation. Gadolinium enhanced T1 cervical MRI, in axial (a), coronal (b) and sagittal (c) shows right occipital condyle mass, with partial extension to the neck and significant homogeneous enhancement. CT shows expansive and osteolytic lesion of the condyle in sagittal image with partial commitment of jugular foramen (d). Positive biopsy for Langerhans cell histiocytosis Group 3.
Atlantoaxial rotatory subluxation

While atlantoaxial rotatory dislocation has been described at all ages; it is pathology almost exclusive to children. It is caused by a flexion mechanism and rotation of the cervical spine associated to transverse ligament weakness, frequently secondary to banal traumas such as minor gymnastic exercises it may also be seen in severe trauma. It is also described in the course of upper respiratory infections (Grisel Syndrome) or post operative tonsillectomy. Clinically, the head is rotated to one side with neck in the opposite direction. The head rotation to the opposite side is limited and accompanied by intense SCM muscle spasm, being impossible to recover the normal position voluntarily or with force. At this height, the spinal canal is large so that this alteration is generally not associated with neurological signs. In simple radiography misalignment of the atlas lateral masses can be seen, one of which is situated in front of the odontoid, a finding that is best represented in CT. Treatment is conservative, consisting of pain management and halter cervical traction. Surgical reduction is rare, although it is described for persistent or recurrent dislocations (Figure 12).

Congenital scoliosis

Scoliosis is defined as the presence of one or more lateral curves of the spine in the coronal plane. It is usually classified as primary (idiopathic) and secondary, which in turn are classified according to its cause. Amongst the causes of scoliosis can be considered neuromuscular diseases, congenital and developmental abnormalities and some tumors. Generally, idiopathic scoliosis is the most common type (80%) followed by congenital scoliosis (10%)\(^1\). Simple radiography and CT are the methods of choice to document bone alterations and MRI is indicated when neuropathic disorder is suspected (Figure 13).
Figura 13. Paciente de 7 meses, tortícolis sin respuesta al tratamiento cinético. Radiografía simple proyecciones frontal (a) y lateral (b) muestran malformaciones vertebrales consistentes en bloques y hemivértebras, mejor demostradas en TC con falta de formación de los arcos anterior y posterior de C1 en reconstrucciones sagital (c) y volumétrica (d).

Conclusion
The spectrum of causes of torticollis in the pediatric age is very broad, and images can be useful for the clinician to achieve an early and accurate diagnosis.

Bibliography