

Bilateral Cervical Ribs Fusing or Naffziger Syndrome: About an Observation in Guinea and Literature Review

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Abstract

Thoraco-brachial outlet syndrome (TDS) or cervical rib brings together all the clinical symptoms (arterial, neurological and/or venous) due to extrinsic, intermittent or permanent compression of the vascular-nervous elements of the root of the upper limb during cervico-thoraco-brachial crossing syndrome. This pathology is relatively common but often overlooked; it affects women more often (between 4/1 and 2/1) and is rare in children. We report the case of a 59-year-old female patient seen in the department for paresthesia and muscular weakness of the upper limbs rated 3/5 on the right and 4/5 on the left progressively evolving over 39 years, electromyography (EMG) showed indicated compression of the brachial plexus and the CT scan confirmed the presence of bilateral cervical ribs fused with the first on the left. She benefited from physiotherapy while awaiting her surgery, and unfortunately, she died of a stroke in the traumatology department. This pathology is the first in our department to be supported by a review of the literature.

Keywords

Cervical Ribs, Naffziger Syndrome, Guinea

1. Introduction

Thoraco-brachial outlet syndrome (TDS) or cervical rib includes all clinical symptoms (arterial, neurological and/or venous) due to extrinsic, intermittent or per-

manent compression of the vasculo-nervous elements of the root of the upper limb during the cervico-thoraco-brachial crossing [1].

This pathology is relatively common but often unknown, it affects women more often (between 4/1 and 2/1) and is rare in children [2].

The first treatments date back to the 1950s [2], however, the first descriptions of positional compression in the shoulder “scale-nus anticus syndrome” date back to the 19th century and Adson’s work (or even the homonymous maneuver) dates back to the very beginning of the 20th century [3].

In the majority of cases, the clinical manifestations are dominated by subjective complaints, which are difficult to attribute with certainty to proximal nerve compression in the thoracobrachial crossing [3].

We report a case of an adult patient in our department and will support it with a review of the literature.

The objective was to present the first observation of the thoraco-brachial outlet (TDBC) or cervical rib to support it with the literature review.

2. Observation

This is a 59-year-old patient admitted to the department for progressive paresthesia and muscle weakness of the upper limbs rated at 3/5 on the right and 4/5 on the left, progressing to progressive worsening since the age of 20.

Clinical examination reveals a sensory motor deficit of the upper limbs, muscular atrophy, a claw-like appearance of the hands (**Figure 1**), as well as a decrease in the axillary pulse indicating a thoracic outlet syndrome.

At the paraclinical level, electromyography (EMG) indicated compression of the brachial plexus predominant on the lower trunk without root rupture.

CT scan confirmed the presence of the bilateral cervical dimensions fused with the first one on the left (**Figure 2**).

Faced with these clinical and paraclinical arguments, the thoracic brachial outlet syndrome (TDBC) or fused cervical rib was put forward and the patient was treated by physiotherapy with slight improvements (muscle weakness in the upper limbs rated at 4/5 on both sides) but unfortunately died as a result of a cerebrovascular accident (CVA) before surgery could be considered in the traumatology department where she was transferred.



Figure 1. Hands of the 59-year-old patient with muscular atrophy and claw-like appearance.



Figure 2. Computed tomography showing bilateral cervical dimensions fusing with the first on the left in the 59-year-old patient followed in the Rheumatology Department of the Ignace Deen National Hospital in Conakry.

3. Discussion

Our observation is that of a thoracic brachial outlet syndrome (TDBC) or fused cervical rib, which is of epidemiological, diagnostic, therapeutic and prognostic interest.

It is relatively rare to be discovered in 1.5% of the general population [4], but even rarer when it occurs bilaterally and fuses with the first, as reported in our observation. This is called “neck ribs”.

Our observation concerns an adult patient with bilateral supernumerary ratings. In the literature, the presence of supernumerary odds is rarely found because it is in 9 out of 10 asymptomatic cases. In a study carried out on deficient forms of thoracobrachial crossing syndrome with 53 cases, including 41 women, with a mean age of 39 years with extremes of 9 - 80 years, the cervical rib was found in 10 cases over a period of 25 years without fusion with the first ribs [3]. Pediatric forms were observed in the same study in 2 cases with isolated sensory deficits in the territory of the medial antebrachial cutaneous nerve (NCABM) [4]. There are few publications in the literature concerning these neurological deficit forms [5]. It seems useful to us to report our observation, which is the first in our department, and to support it with a review of the literature based on the epidemiological, diagnostic, therapeutic and prognostic interests.

These forms are rare, their prevalence being estimated at one case per million inhabitants, so their diagnosis is often delayed [3] [6]. In our patient, it was based on the clinical signs of sensorymotor deficit of the upper limbs with muscular atrophy, a claw-like appearance of the hands associated with the paraclinical signs (X-ray and CT scan) which showed fusing bilateral cervical ribs, the EMG had highlighted the tracings in favor of compression of the brachial plexus predominant on the lower trunk without root rupture. In the literature, the topography of clinical signs and ENMG results show that they are always related to damage to the lower trunk or the medial bundle of the brachial plexus [7]. Motor deficit

involves intrinsic muscles with muscular atrophy that begin with the lateral thenar muscles and then affects the interosseous and hypothenar muscles with a claw deformity of all long fingers [7] [8].

In these deficient forms, bone abnormalities, such as a cervical rib or apophyseomegaly, are often present [9].

The resting clinical examination is most often normal and the symptoms are detected during the performance of dynamic maneuvers, which will be detailed below. The most reported symptoms are the presence of pain following the mobilization of the upper limb, particularly during the performance of abduction movements, as well as the presence of muscle fatigability. These symptoms are only slightly predictive of the type of vascular or neurological involvement [10] [11]. It is important to remember that there are no pathognomonic signs of an attack. The neurological signs are rather cramp-like, with heavy pain located in the cervical and shoulder if affected by C5-C6 roots radiating into the occipital region. In the case of C8-D1 low root damage, the symptoms follow the path of the ulnar nerve. It can also correspond to functional impotence, the presence of elective paresthesia of the last two fingers.

In a number of patients, the clinical symptoms are not very specific, and the diagnosis is actually made based on the recurrence of the patient's symptoms over various periods.

Dynamic maneuvers (72% sensitivity, 53% specificity) [12]-[14], including:

Adson's Maneuver: Inspiration and Rotation-Ipsilateral Neck Extension, Positive Upon Pulse Abolition and/or Awakening of Clinical Symptomatology (Sensitivity: 79%, Specificity: 76%) [15] [16].

Wright's manoeuvre: arm in anatomical position, abduction elevation at 30, 60, 90 and 180° with measurement of the abduction angle for which the pulse disappears (Specificity: 70%, Sensitivity: 53%) and the symptomatology of the seated patient reappears [17].

Tinel's maneuver: digital pressure and percussion of the supraclavicular and subclavicular hollows aimed at reproducing the clinical symptomatology (sensitivity: 46%, specificity: 56%) and the Roos test: elevated position abduction of the arms, bent elbows, with alternating flexion-extension of the fingers for at least three minutes, aimed at reproducing the clinical symptomatology (Sensitivity: 84%, Specificity: 30%) [18] [19].

The treatment is based on rehabilitation aimed at correcting the dysfunction of the scapular musculature, relaxation of the cervicospinal region, and isometric strengthening of the muscles (trapezius, scapula elevator, sternocleidomastoid and serratus anterior). Physical therapy sessions should be accompanied by daily self-rehabilitation exercises [20] [21].

It is only surgical in the event of failure of rehabilitation or severe symptoms: scalenotomy.

Resection of a cervical rib or the first rib, section of a fibrous band, thromboembolism, arterial reconstruction.

The surgical indication must be made according to the predominant sympto-

matology, the various investigations, and after 6 months of well-conducted physiotherapy. The surgical strategy aims to decompress the vasculonervous structures in the thoracobrachial crossing. It includes the section of the anterior and middle scalene muscles, and the excision of the first rib (and section of the pectoralis minor, only in the case of additional compression). The technique is that of first rib resection, whether or not associated with surgery of the pectoralis minor, and which can be performed in two modalities: supraclavicular or Roos' axillary approach [20] [21].

The transaxillary approach: patient in a lateral position with traction of the arm at the zenith to open the costoclavicular space. The vasculo-nerve structures are pulled upwards and cleared to allow the complete removal of the first rib, from "cartilage to cartilage": from the transverse process (posterior joint) to the costochondral junction (anterior joint). This transaxillary approach is reputed to be more difficult. The narrow opening and difficulty of controlling vascular structures by this route have contributed to its bad reputation. However, it allows for satisfactory C8-T1 neurolysis, complete excision of the first rib, and an extra cervical rib if present. The scar in the hollow of the armpit has an aesthetic advantage, but a risk of longer and more complicated healing because it is in a maceration area.

The supraclavicular approach is the preferred route when vascular reconstruction is necessary. The patient is in the supine position. The incision is parallel to the collarbone. This approach allows good exposure of the scalene muscles but exposes the risk of damage to the phrenic nerve due to its path to the anterior aspect of the scalene muscles. The principle remains the same with the section of the scalene muscles and excision of the first rib. In rare cases, such as isolated compression by the pectoralis minor muscle, an isolated approach with regard to the coracoid process may be performed to section the pectoralis minor muscle. In the event of additional compression, the transaxillary approach allows it to be crossed. Procedures associated with first rib resection may be necessary: treatment of a fibromuscular anomaly, arterial or venous reconstruction, thoracic sympathectomy. The main complications of surgery are vascular and neurological wounds due to damage to the brachial plexus or phrenic nerve and hemothorax. There is a clear improvement in vascular forms, which is less important for nervous forms.

The functional prognosis and the resumption of sport are largely conditioned by the presence or absence of vascular or nervous complications at the time of diagnosis, which from our point of view justifies a rather clear attitude in favor of in-situ fibrinolysis in the forms revealed by an initial thrombophlebitis. After surgery, the resumption of gentle and then active passive mobilization, avoiding abduction movements and load-bearing, must be early and last at least one month before any resumption of fuller or more sustained activity [22]. Resumption of the sporting gesture is sometimes proposed around the second postoperative month without forcing the pace [23]-[24].

The choice of approach may be dictated by operational constraints and depends on the surgical schools and experiences.

4. Conclusions

Thoracic outlet syndrome due to supernumerary ribs is a rare pathology and difficult to diagnose. However, a good clinical examination and a careful examination can guide the diagnosis (see **Figure 3** and **Figure 4**).

The neurological deficit is slow-onset, delaying the consultation because the patient only notices frank muscular atrophy after several years of evolution. Sometimes, there is an old muscular atrophy of the lateral thenarians, but the patient consults a doctor when the patient has recently appeared with a claw-like deformity of the fingers.

Its management depends on the symptomatology, a priori rehabilitation, physiotherapy or surgery which allows a disappearance of the pain and a stabilization or at least a partial recovery of the deficit which improves the functional and vital prognosis.

Perspective think about it in the face of the sensorimotor deficit of the upper limbs with muscular atrophy and a claw-like appearance of the hands.

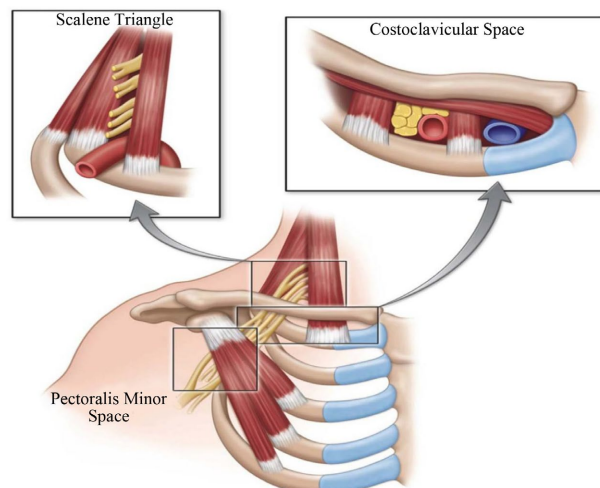


Figure 3. To facilitate the assimilation of this pathology, we will provide a diagram. This diagram are represented the three potential sites of compression within the cervicoaxillary canal. [25] [26]

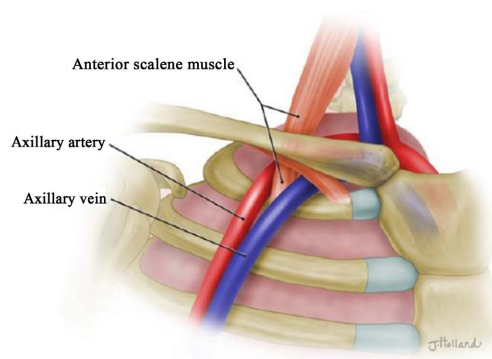


Figure 4. Likewise the basic anatomy of the thoracic outlet, the vein moving anterolateral relative to the artery and in front of the anterior scalene muscle. [27]

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Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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