

## CASE REPORT

# Spontaneous cervical swelling syndrome as a rare cause of neck edema: case series and literature review

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## Abstract

Spontaneous cervical swelling syndrome (SCSS) is a rare disorder characterized by unprovoked, self-limiting and often unilateral cervical edema. SCSS is a recurrent disorder that predominantly affects adult women and is not associated with laboratory abnormalities. We report on eight female patients with a mean age of 56 (38–82) years at the time of the first presentation. The episodes were characterized by an acute onset in all patients and had a mean duration of 3.8 (1–7) days. Biochemical analysis did not reveal any related abnormalities. Imaging of the neck and chest demonstrated diffuse edema in the supraclavicular fossa and left infrahyoid region in all patients. At the time of the acute event, lymphatic scintigraphy revealed tracer accumulation in the left supraclavicular region in three patients and could not demonstrate any abnormalities in the in-between episodes in two patients.

## INTRODUCTION

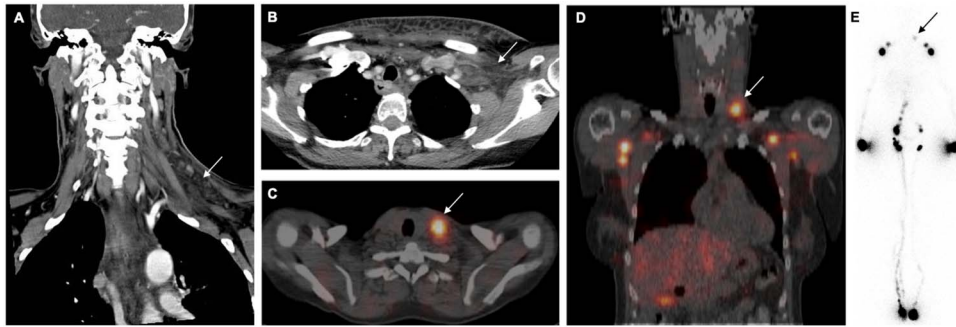
In the past, several terms have been used to refer to patients with spontaneous, atraumatic swelling of the left supraclavicular fossa, including 'benign supraclavicular tumorous lymphangiectasia' and 'recurrent lymphangiectasia of the left supraclavicular fossa'. Franceschi *et al.* were the first to report on this syndrome as a distinct clinical entity [1]. The pathogenesis remains to be fully elucidated, but a transient obstruction of

the thoracic duct is suspected to be the cause, which may account for the extravasation of chyle during the acute event and normalization of lymphatic transport in the in-between episodes. Over the past few years, eight patients consulted our tertiary care facility because of a spontaneous, atraumatic swelling of the left supraclavicular fossa. In this case series, we report on the epidemiology, clinical presentation and imaging findings in the spontaneous cervical swelling syndrome (SCSS).

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**Figure 1:** (A) Coronal contrast-enhanced CT image showing diffuse left-sided edema in the base of the neck (arrow) extending to the upper axilla. (B) Axial contrast-enhanced CT image showing diffuse left-sided edema in the base of the neck, upper axilla and anterior chest wall (arrow). The edema crosses the midline in the anterior chest wall. Mediastinal edema is noted, causing some deviation of the trachea and esophagus. (C) Axial and (D) coronal  $^{99m}\text{Tc}$ -Nanocolloid SPECT/CT fusion images showing tracer accumulation in the left cervical region (arrow). (E) Whole-body  $^{99m}\text{Tc}$ -Nanocolloid lymphatic scintigraphy showing lymphatic extravasation in the left supraclavicular fossa (arrow).  $^{99m}\text{Tc}$ , technetium-99m.

## CASE SERIES

All patients ( $n=8$ ) were female with a mean age of 56 (38–82) at presentation. Five out of 8 patients were perimenopausal or menopausal at the time of diagnosis. All episodes had an acute onset of the supraclavicular fossa swelling. Seven patients reported having multiple episodes in the past. The mean duration of episodes in patients with symptom-free intervals was 3.8 [1–7] days. Physical activity, including jogging and gardening, and warm weather conditions were reported as eliciting factors. The most frequently reported associated symptoms include a cervical pressure sensation, shortness of breath and general malaise. Clinical examination revealed left-sided, non-tender and non-pitting edema of the supraclavicular fossa in all patients. The edema extended to the left cervical region over the course of the episode in seven patients. The clinical characteristics are summarized in Table 1.

The laboratory results did not reveal abnormalities in six patients. Patient 1 had a minor elevation of D-dimers and Patient 4 had long-standing anemia, which were considered not to be related to the cervical edema (Table 2). Ultrasound with echocolor Doppler revealed subcutaneous edema in all patients and thoracic duct dilatation in four patients. Computed tomography (CT) of the neck and chest showed edema of the supraclavicular fossa in all patients (Fig. 1A and B), with the edema extending to the left cervical region in seven. In three patients, retropharyngeal edema extending up to level C2 was noted. The larynx, thyroid gland, proximal trachea and esophagus were slightly deviated to the right in seven patients. In two patients, the cervical part of the thoracic duct appeared to be wide. Furthermore, we noted lymphadenopathy in all patients, mediastinal involvement in seven patients and pleural fluid in five patients. Ascites was present in one patient. All vascular structures appeared patent, there was no evidence of a thrombosis. Single-photon-emission computed tomography (SPECT)/CT lymphoscintigraphy was performed in five patients, with three out of five scans performed during the acute episode (Fig. 1C–E). In all three scans performed at the time of the acute event, tracer accumulation was visualized in the left cervical region. In two patients, SPECT/CT lymphoscintigraphy did not reveal any pathological findings after the disappearance of the cervical swelling, 2 and 3 weeks after the onset of the episode, respectively.

## DISCUSSION

In opposite of the term proposed by Franceschi *et al.*, we prefer the term ‘SCSS’, as in some patients the edema may be non-recurrent or persistent in nature. Furthermore, it emphasizes the unprovoked nature of the SCSS. All patients included in this case series are female, with the majority being either perimenopausal or menopausal. Preyer *et al.* suggested the possible influence of external estrogen administration, considering all their patients were treated with estrogen supplements respectively [2]. Among these patients, only one received estrogen supplements. Physical activity and warm weather conditions were reported to be possible triggers. Other authors also have reported strenuous exercise and household chores as possible precipitating factors [2, 3].

Based on our case findings and the literature review (Supplementary data), we propose a case definition for SCSS (Table 3). However, it remains challenging to exclude other possible causes of acute cervical swelling (Table 4). A subacute onset without spontaneous regression should raise the suspicion for a tumoral process [4–7]. Systemic capillary leak syndrome should be suspected in case of hypotension, hemoconcentration and hypoalbuminemia. Hereditary angioedema may also present with recurrent swelling, although usually bilateral, and can be excluded by measuring the C1 esterase inhibitor activity. Fever and elevation of inflammatory parameters should raise suspicion for an underlying infectious or systemic inflammatory disease. Cytopenia and serum protein electrophoresis abnormalities may indicate a hematologic malignancy. D-dimers can be useful in patients with suspected thrombosis, although this should be actively excluded with imaging. A post-traumatic origin swelling should be actively questioned.

Imaging may aid in excluding the underlying structural abnormalities. While ultrasound may be indicative of the SCSS by showing edema and thoracic duct distension, it is highly user-dependent and often inadequate to rule out other local and systemic causes of cervical swelling. Two of our patients had false-positive ultrasound results in which the swelling was initially attributed to a multinodular goiter and a cystic lesion, respectively, that could not be confirmed on a subsequent CT scan. The extension of the lymphedema and presence of pleural fluid, mediastinal involvement and lymphadenopathy may also be evident on a CT of the neck and chest. In one of

Table 1: Clinical characteristics of our patient cohort

|           | Gender | Age at first presentation (years) | Medical history   | Menopausal state | Episodes   | Duration (days) | Triggers                             | Associated symptoms                |
|-----------|--------|-----------------------------------|---|------------------|------------|-----------------|--------------------------------------|------------------------------------|
| Patient 1 | Female | 58                                | Allergic rhinoconjunctivitis                                  | Menopausal       | 2          | 2               | None                                 | Pleuritic pain<br>Dyspnea          |
| Patient 2 | Female | 46                                | None  | Not reported     | 1          | 4-5             | Physical exercise                    | None                               |
| Patient 3 | Female | 65                                | Hypertension<br>Monoclonal gammopathy of unknown significance | Menopausal       | 3          | 3               | Physical labor<br>Vaccination (DTaP) | Pressure sensation<br>Dyspnea      |
| Patient 4 | Female | 58                                | Breast cysts<br>Ovary cysts                                   | Menopausal       | Recurrent  | 1-7             | Physical exercise<br>Physical labor  | Pleuritic pain<br>Dyspnea<br>Cough |
| Patient 5 | Female | 53                                | Nasal polyposis   | Perimenopausal   | Recurrent  | 3-4             | Warm weather                         | Pressure sensation<br>Malaise      |
| Patient 6 | Female | 38                                | Allergic rhinoconjunctivitis                                  | Pre-menopausal   | Persistent | Persistent      | None                                 | None                               |
| Patient 7 | Female | 47                                | De Quervain thyroiditis<br>Lymphomatoid papulosis             | Not reported     | 2          | 7               | None                                 | Pleuritic pain<br>Abdominal pain   |
| Patient 8 | Female | 82                                | Parkinson disease   | Menopausal       | Recurrent  | 2               | Warm weather                         | Dyspna<br>Malaise                  |

Abbreviations: DTaP, diphtheria, tetanus acellular pertussis.

Table 2: Results of laboratory examination and imaging techniques

| Patient   | Laboratory abnormalities                 | US/CT neck findings   |                     |                         | CT thorax       |  | SPECT lymphoscintigraphy |
|-----------|--|---|---------------------|-------------------------|-----------------|--|--------------------------|
|           |  | US/CT neck findings   | Pleural fluid       | Mediastinal involvement | Lymphadenopathy |  |                          |
| Patient 1 | D-dimers (570 ng/ml; normal < 550 ng/ml) | Edema of the supraclavicular fossa and left cervical region extending toward the paratracheal region—thoracic duct distension absent  | Present (left)      | Present                 | Present         | Not performed  |                          |
| Patient 2 | None                                     | Edema of the supraclavicular fossa—thoracic duct distension present   | Not documented      | Present                 | Present         | Not performed  |                          |
| Patient 3 | None                                     | Edema of the supraclavicular fossa and left cervical region extending toward the paratracheal region—thoracic duct distension absent  | Present (bilateral) | Present                 | Present         | Extravasation in both axillary regions and the left supraclavicular region |                          |
| Patient 4 | Long-standing anemia                     | Edema of the supraclavicular fossa and left cervical region—thoracic duct distension absent   | Present (bilateral) | Present                 | Present         | Normal captation (3 weeks after onset of episode)                          |                          |
| Patient 5 | None                                     | Edema of the supraclavicular fossa and left cervical region extending toward the paratracheal region—thoracic duct distension present | Present (bilateral) | Present                 | Present         | Normal captation (2 weeks after onset of episode)                          |                          |
| Patient 6 | None                                     | Edema of the supraclavicular fossa and left cervical region—thoracic duct distension absent   | Absent              | Absent                  | Present         | Extravasation left cervical region   |                          |
| Patient 7 | None                                     | Edema of the supraclavicular fossa and left cervical region extending toward the paratracheal region—thoracic duct distension present | Present (bilateral) | Present                 | Present         | Extravasation left cervical region   |                          |
| Patient 8 | None                                     | Edema of the supraclavicular fossa and left cervical region—thoracic duct distension present  | Not documented      | Present                 | Present         | Not performed  |                          |

Abbreviation: US, ultrasound.

**Table 3:** Case definition of the SCSS

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|---|
| Documented soft swelling of the left supraclavicular fossa (±cervical region) on clinical examination   |
| Acute onset (less than 24 h)  |
| Regression over the course of 1 week or less  |
| Recurrence possible   |
| No abnormalities in in-between episodes   |
| Normal laboratory analysis (absence of raised inflammatory markers, cytopenia, serum protein electrophoresis abnormalities, hemoconcentration, hypoalbuminemia and C1 esterase deficiency, ±D-dimers) |
| Imaging demonstrates edema of the supraclavicular fossa and left cervical region  |

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**Table 4:** Differential diagnosis of acute and subacute cervical swelling

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- Malignant neck tumors
    - Primary solid or non-solid tumor
      - Lymphoma
      - Osteosarcoma
    - Metastatic disease (Virchow's node)
  - Infectious disease
    - Abscess
    - Cellulitis
    - Lymphadenitis
  - Vascular disorders
    - Thrombosis of subclavian or internal jugular vein
    - Internal jugular vein phlebectasia
    - Arterial dissection
    - Superior vena cava syndrome
  - Systemic and inflammatory disease
    - Kikuchi-Fujimoto disease
    - Systemic capillary leak syndrome
  - Allergic and non-allergic angio-edema
  - Post-traumatic (edema, hemorrhage, external compression)
- 

the patients, who reported abdominal pain associated with the cervical edema, a CT scan of the abdomen confirmed the presence of ascites. Tracer accumulation was visualized in the left supraclavicular region in all patients who underwent lymphatic scintigraphy during the acute episode. By contrast, two patients undergoing scintigraphy between episodes did not demonstrate any abnormalities in accordance with the intermittent nature of this disorder. The transient nature of the SCSS may interfere with the timely performance of appropriate imaging techniques.

In conclusion, SCSS is an uncommon and relatively unknown entity that predominantly affects adult women and causes acute, self-limiting swelling of the left supraclavicular region. SCSS may recur and is not associated with laboratory abnormalities. Lymphatic scintigraphy at the time of the acute episode of cervical edema reveals tracer accumulation in the left supraclavicular region.

## SUPPLEMENTARY MATERIAL

Supplementary material is available at the *Journal of Surgical Case Reports* online.

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## CONFLICT OF INTEREST STATEMENT

None declared.

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## ETHICAL APPROVAL

This study was approved by the Ethical Committee Research UZ/KU Leuven (Belgium).

## INFORMED CONSENT

Written informed consent was obtained from all individuals.

## GUARANTOR

Steven Vanderschueren.

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