CASE REPORT

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

Albrecht Betrains1,2,*, Robert Hermans3,4, Olivier Gheysens4,5, Vincent Vander Poorten6,7, Willy E. Peetermans2 and Steven Vanderschueren2

1Department of General Internal Medicine, University Hospitals Leuven, Leuven, Belgium, 2Laboratory of Clinical Infectious and Inflammatory disease, Department of Microbiology, Immunology and Transplant medicine, University Hospitals Leuven, Leuven, Belgium, 3Department of Radiology, University Hospitals Leuven, Leuven, Belgium, 4Department of Imaging and Pathology, University Hospitals Leuven, Leuven, Belgium, 5Nuclear medicine and Molecular Imaging, University Hospitals Leuven, Leuven, Belgium, 6Department of Otorhinolaryngology, University Hospitals Leuven, Leuven, Belgium, 7Department of Oncology, University Hospitals Leuven, Leuven, Belgium

*Correspondence address. Department of General Internal Medicine, University Hospitals Leuven, Herestraat 49, Leuven 3000, Belgium.
Tel: +32-16344275; E-mail: albrecht.betrains@uzleuven.be

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

Abbreviations: DTaP, diphtheria, tetanus acellular pertussis.
| Laboratory abnormalities | US/CT neck findings | CT thorax | SPECT lymphoscintigraphy |
|--------------------------|----------------------|-----------|--------------------------|
|                          |                      | 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 Normal captation (3 weeks after onset of episode) |
| Patient 4                | Long-standing anemia | Edema of the supraclavicular fossa and left cervical region—thoracic duct distension absent | Present (bilateral) | Present | Present | Normal captation (2 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 | |
| Patient 6                | None                 | Edema of the supraclavicular fossa and left cervical region—thoracic duct distension present | 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.
Spontaneous cervical swelling syndrome

Table 3: Case definition of the SCSS

| Definition                                                                 |
|----------------------------------------------------------------------------|
| 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, hypoalbuminemia and C1 esterase deficiency, ±D-dimers) |
| Imaging demonstrates edema of the supraclavicular fossa and left cervical region |

Table 4: Differential diagnosis of acute and subacute cervical swelling

- 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)

SUPPLEMENTARY MATERIAL

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

ACKNOWLEDGEMENTS

We would like to thank Barbara Cassimon, MD, for her help with an earlier version of the manuscript.

CONFLICT OF INTEREST STATEMENT

None declared.

FUNDING

This research did not receive any specific grant from funding agencies in the public, commercial or not-for-profit sectors.

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.

REFERENCES

1. Franceschi C, Gianesini S, Bahnini A, Laurian C, Menegatti E, Zamboni P. The recurrent cervical swelling syndrome. Phlebology 2012;27:90–2.
2. Preyer S, Kaiserling E, Heinle H, Foldi E, Zenner H, Foldi M. Benign supraclavicular tumorous lymphangiectasia—a new disease? Lymphology 1995;28:118–25.
3. Suzuki M, Morita S, Iizuka K. A case of idiopathic lymph leakage in the neck. Auris Nasus Larynx 2010;37:535–7.
4. DePena C, Van P, Lee Y. Lymphoma of the head and neck. Radiol Clin North Am 1990;28:723–43.
5. Wax M, Yun K, Omar R. Extramedullary plasmacytomas of the head and neck. Otolaryngol Neck Surg 1993;109:877–85.
6. Wanebo H, Koness R, Macfarlane J, Eilber F, Byers R, Elias E, et al. Head and neck sarcoma: report of the head and neck sarcoma registry. Head Neck 1992;14:1–7.
7. Cervin J, Silverman J, Loggie B, Geisinger K. Virchow’s node revisited. Analysis with clinicopathologic correlation of 152 fine-needle aspiration biopsies of supraclavicular lymph nodes. Arch Pathol Lab Med 1995;119:727–30.