Bilateral axillary web syndrome in a patient with primary lymphoedema of upper limbs and non-Hodgkin lymphoma

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Abstract

Primary lymphoedema, axillary web syndrome (AWS) and yellow nail syndrome may be related. Mr B is a 66-year-old gentleman with genital lymphoedema and lymphoedema of all four extremities. In 2023, he was diagnosed with non-Hodgkin lymphoma and also underwent cardiac surgery. In November 2023, he completed an inpatient rehabilitation at the Földi clinic in Germany, where he received intensive treatment for his lymphoedema and was also diagnosed with bilateral AWS. The presence of AWS in a patient with primary lymphoedema and no history of axillary surgery is unique. Although AWS typically presents after axillary surgery, this case highlights that it can also occur in patients without lymph node surgery. While the precise cause of this presentation of AWS is not known, it may be connected to yellow nail syndrome or potentially the recent chemotherapy treatment. This article will describe the clinical case, highlighting the need for further research on AWS present in primary lymphoedema.

Keywords: axillary web syndrome • primary lymphoedema • physiotherapy • yellow nail syndrome

Axillary web syndrome (AWS) refers to the development of thin 'guitar string-like' fibrotic bands or cords in the axilla of patients. The primary cause of AWS is widely recognised as being a direct result of axillary lymph node dissection for breast cancer (Yeung et al, 2015; Dinas et al, 2019; Gonzalez-Rubino et al, 2023). However, some cases have been reported in which axillary ganglionar emptying was not the cause of AWS. Welsh and Gryfe (2016) discussed a rare case of AWS following a period of intense sport. They reported that the 38-year-old competitive squash player developed the cords approximately 24 hours after a particularly aggressive game of squash. He was treated with manual therapy and self-massage.

Lymphoedema is the swelling of one or several parts of the body because of accumulation of lymph in the extracellular space (Földi, 2015). It is a chronic condition which may worsen if untreated, predisposes the body to infections and causes a reduction in quality of life. The chronic, progressive accumulation of protein-rich fluid in the interstitium exceeds the capacity of the lymphatic system to transport the fluid (Sleigh and Manna, 2022). Swelling associated with lymphoedema can occur anywhere in the body, including the arms, legs, genitals, face, neck, chest wall and oral cavity (Hasenoehrl et al, 2020; Brouillard et al, 2021; Sleigh and Manna, 2022).

Yellow nail syndrome is a rare condition defined by a presence of two of the following: slow-growing, hard, yellow and dystrophic nails; lymphoedema; and respiratory tract disease (Cheslock and Harrington, 2023; Walz-Eschenlohr et al, 2023). While the aetiology of AWS does not influence treatment, more studies offering treatment for non-surgical aetiology would be of interest (Yeung et al, 2015).

Full informed, written consent was obtained from the patient when the team decided to initiate the case report,

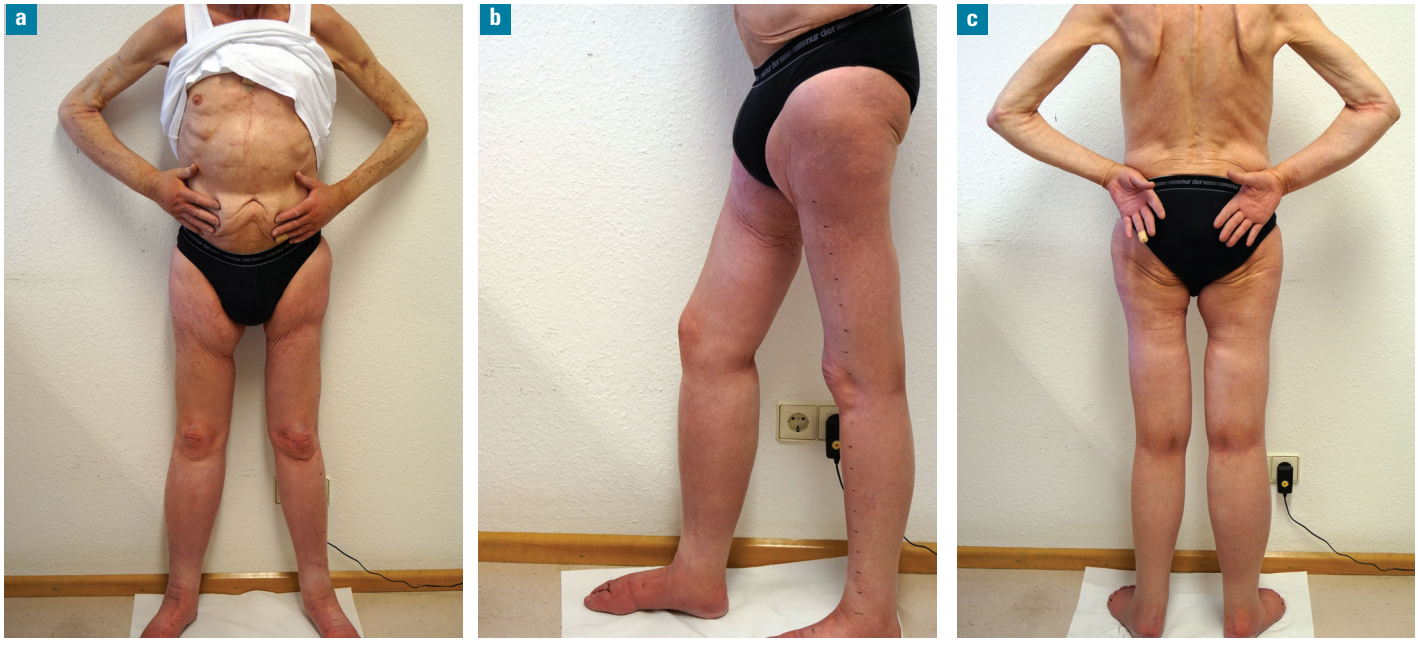

Figure 1. Mr B on presentation in October 2023

including further publication related to his treatment. For the preparation of this case report, the 2013 CARE Checklist has been taken into account (CARE Case Report Guidelines, 2013).

Case description

Mr B is a 66-year-old retired government worker who lives with his wife. He has lymphoedema in all four limbs, as well as genital lymphoedema due to protein-losing enteropathy with chylous pleural effusions and chylous ascites (*Figure 1*). He presented with yellow nail syndrome (*Figure 2*).

In February 2023, he was diagnosed with non-Hodgkin lymphoma and was treated with five cycles of chemotherapy. The sixth cycle could not be administered as a result of bilateral pneumonia in the context of leukopenia. He has had recurrent erysipelas, the most recent case was on his right forearm, which was reported in August 2023. Mr B was known to the team after he had had aortocoronary bypass surgery in 2002 for coronary artery disease. He had also been treated for chronic obstructive pulmonary disease and sleep apnoea syndrome.

On assessment, no enlarged lymph nodes were palpable. There was generalised oedema, with only the head, neck and shoulders being free of oedema. He presented with four cords in the left axilla (*Figure 3*), ranging from the internal sulcus of the pectoralis major to the forearm, and two cords in the right axilla (*Figure 4*) to the middle of the arm. They compromised shoulder movement in the last degrees of abduction and flexion, but functionality was not particularly impaired. Until November 2023, he had not noticed the presence of bilateral AWS. It was diagnosed during his rehabilitation stay at the Földi clinic. For better visualisation and palpation of the AWS, the arm was placed in abduction and flexion of the shoulder, elbow in extension and ulnar deviation of the wrist (Yeung et al, 2015). In this way, the lymphatic cords were put under maximum tension. No lymphatic chords were found in the groin area.

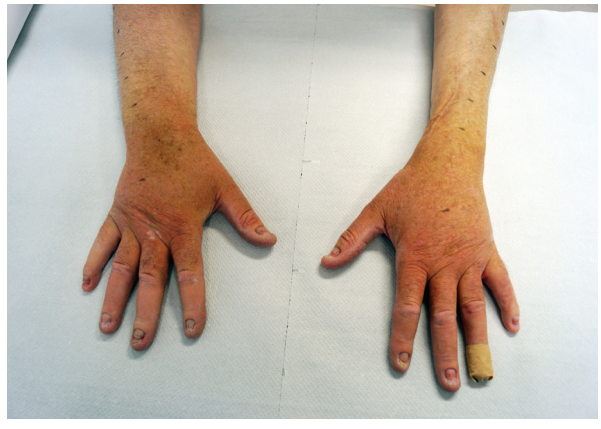

Figure 2. Yellow nail syndrome is evident on Mr B's hands

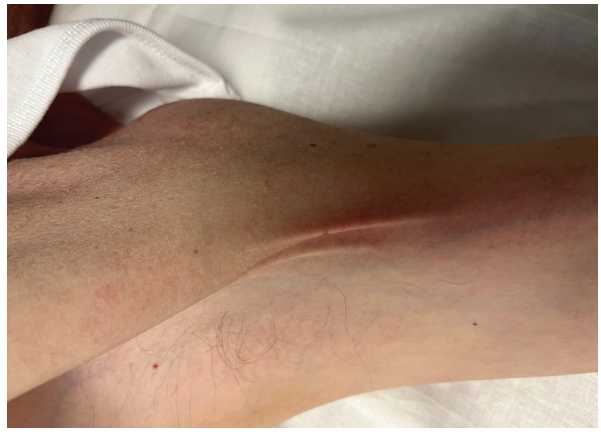

Figure 3. Axillary web syndrome seen in the left axilla

The AWS slightly limited Mr B's last degrees of shoulder movement with pain present on palpation, scoring 3 out

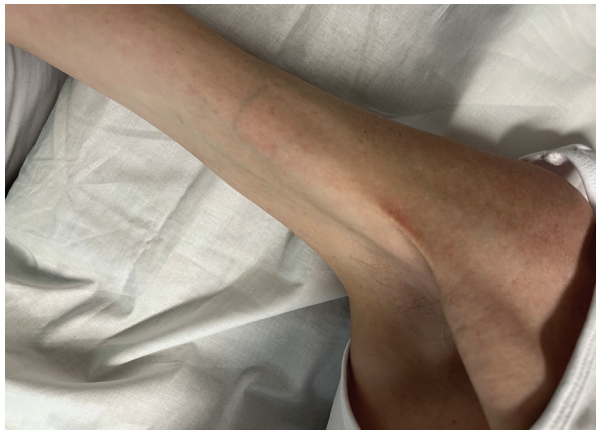

Figure 4. Axillary web syndrome seen in the right axilla

of 10 on a visual analogue scale. According to the Global Physical Activity Questionnaire (GPAQ) scale, the patient had a moderate level of physical activity capacity (Keating et al, 2019). Results from the Disabilities of the Arm, Shoulder and Hand study (Roy et al, 2009) indicated a partially reduced upper limb function. Results from the European Organization for Research and Treatment of Cancer Quality of Life Questionnaire Core 30 (Jurys and Durmala, 2021) showed a minimally reduced quality of life score. When he presented, he had been wearing custom-made compression garments on all four limbs during the day with good tolerance.

Intervention

During his stay at the Földi clinic, Mr B received twice daily lymphatic drainage for all four limbs and compressive bandaging to treat his lymphoedema. AWS treatment involved stretching and non-painful massage of the lymphatic cords in the armpit, arm and forearm. Each AWS treatment session lasted 30 minutes. The physiotherapist placed the patient's shoulder in abduction, with the elbow extended while massaging the lymphatic cords under tension. The massage was applied to the axilla and the AWS pathway in the arm.

Outcomes

Substantial limb volume reductions at the time of discharge were observed and are given in *Table 1* and *Figure 5*. The patient lost perimetric volume of all his extremities, especially his right leg where he saw a 40% reduction in overall limb volume (*Figure 5*).

Since specific treatment of AWS was limited due to time constraints, they did not disappear completely. However, mobility was improved and the tension of the cords decreased when Mr B performed maximum shoulder abduction.

Discussion and implications for practice

Yellow nail syndrome and lymphoedema are often associated, as in the case of this patient—presented with generalised lymphoedema leaving out only the torso (i.e. the back, abdomen, thorax, head and shoulders). The peculiarity of this case is that the patient presents with AWS in both axillae and arms without previous lymph node surgery. It is normally associated with patients who have had some type of lymph node surgery and is very frequent in patients with breast cancer.

The time of evolution of AWS could not be determined precisely as the patient was not aware of having it until he was diagnosed during the course of his rehabilitation treatment. AWS usually presents with limited limb mobility and pain (Yeung et al, 2015). However, the patient had minimal limitation of the functional mobility of his shoulder, at least for activities of daily living. Although he only had minimal volume reduction in his upper limbs, it is possible that the presence of lymphoedema in his arms restricted his capacity to visualise himself and may have hindered the diagnosis.

Since the lymphatic cords present in this patient do not limit mobility much, we assume that they are more elastic and flexible than those that do. Research indicates that they usually disappear with stretching and physical therapy (Yeung et al, 2015; Gonzalez-Rubino et al, 2023). If left untreated, AWS has been reported to commonly disappear in less than a year (Dinas et al, 2019); however, it is also known to still be present 5 years post surgery (Koehler et al, 2022). The ongoing presence of these cords could therefore indicate

Table 1: Volume measures of Mr B from arrival to discharge

Date	Left arm				Right arm			
	Vol. upper	Vol. lower	Limb vol.	Difference	Vol. Upper	Vol. lower	Limb vol.	Difference
04.10.2023	859.26	698.34	1557.6		1254.33	777.87	2032.20	
01.12.2023	823.16	692.97	1516.13	-41.47 (-2.73%)	1190.57	768.08	1958.65	-73.55 (-3.75%)
Date	**Left leg**				**Right leg**			
	Vol. upper	**Vol. lower**	**Limb vol.**	**Difference**	**Vol. Upper**	**Vol. lower**	**Limb vol.**	**Difference**
04.10.2023	4315.33	3842.10	8157.43		5476.19	4348.86	9825.05	
01.12.2023	3349.89	3607.78	6957.67	-1199.76 (-17.24%)	4148.09	4035.75	8183.84	-1641.21 (-20.05%)

either a recent onset, possibly associated with his recent chemotherapy or heart surgery, or perhaps more likely, given neither his cancer treatment nor cardiac intervention was centred on his axillae, the cords are of long evolution and have not spontaneously disappeared with everyday movements. This could highlight a significant difference between the presence of AWS post-breast cancer treatment and AWS in primary lymphoedema.

Limitations

One of the limitations of this case study is the limited treatment time, which was mainly due to restricted inpatient time. If the patient had received more treatment sessions, he may have regained full shoulder mobility, and AWS might have disappeared. In a more general perspective, the study of AWS is limited by the low prevalence of the syndrome precluding the performance of larger studies. Moreover, there is no common agreement regarding the optimal physiotherapeutic approach to the treatment of axillary web syndrome.

The existence of generalised lymphoedema could be associated with enteropathy and its problem of intestinal protein absorption (Vergara de la Campa et al, 2019; Bosch Taltavull et al, 2023). The review by Yeung et al (2015) quotes a study in which four patients experienced recurrence of AWS after receiving chemotherapy. In this clinical case, although the patient did not have axillary lymph node surgery, the chemotherapy treatment may have been the trigger for the AWS.

There are very few publications on this subject and there is a great need to understand this type of primary globalised lymphoedema and its possible relationship with AWS.

Conclusion

Although AWS typically presents post-axillary surgery, this case highlights that it can also occur in patients without lymph node surgery. While the exact cause of this presentation of AWS is not known, it may be connected to yellow nail syndrome or potentially, the recent chemotherapy treatment. There are very few publications addressing the aetiology, pathophysiology and treatment of AWS in patients without previous lymph node surgery. More research focusing on non-surgical-related AWS is needed, which will provide the basis for optimal selection of treatment tools, as well as a deeper understanding as to whether techniques applied in cases of surgically-caused AWS are as effective as those applied in non-surgically caused AWS. BJCN

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Declaration of interest: None

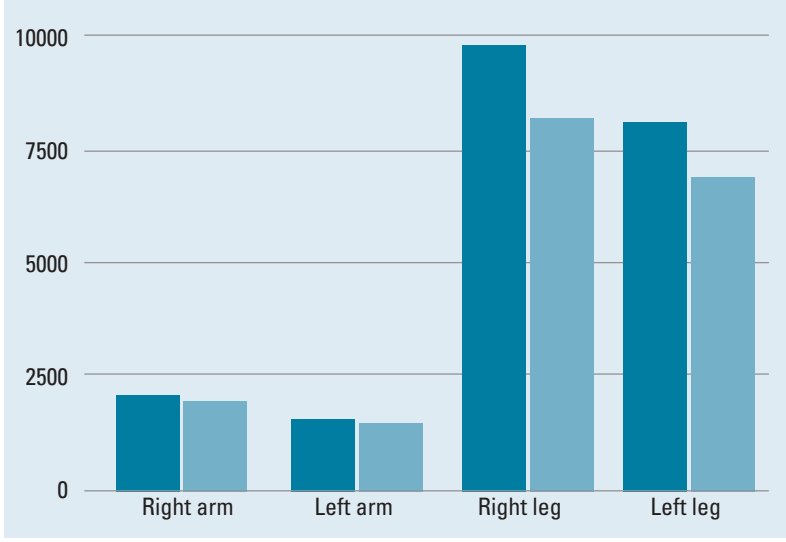

Figure 5. Volume measures of Mr B from arrival to discharge

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