



## Case Report

## Combined vascular malformation of neck: A case report



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## ABSTRACT

Vascular malformations are developmental anomalies occurring due to defective vasculogenesis. Depending on the number of vessels involved, they are subgrouped into simple or combined types. Combined vascular malformations are a rare clinical entity with two or more vascular malformations (capillary, venous, arteriovenous, lymphatic) present in one lesion. Due to the complexity of these lesions, clinicians should employ an interdisciplinary approach with multi-staged treatment for the proper management. Here, we report a case of a lady with a combined vascular malformation in the right side of the neck who presented to our department with a complaint of progressively increasing swelling for three months. A multidisciplinary team of vascular surgeons, interventional radiologists and physiotherapists were involved in its treatment. Initially, Doppler ultrasonography was performed, which revealed a vascular lesion with arterial and venous components and a flow void region suggestive of lymphatic malformation, further confirmed by magnetic resonance imaging (MRI). A multistage treatment modality was employed where intralesional steroid was initially administered in the lesion area to shrink the vessel's size. Next, venous malformation targeted sclerotherapy was performed preoperatively, followed by intraoperative ligation of the feeding vessel and excision of remaining malformation. Postoperatively the patient had no wound site complication and was discharged on the fourth postoperative day with advice to follow rehabilitative neck exercises. One month follow-up revealed complete resolution of the malformation. Thus, in the case of combined vascular malformation, multistage treatment modalities with a multidisciplinary team should be employed for proper treatment.

## 1. Introduction

A combined vascular malformation involving all three components—artery, vein, and lymphatic vessels—is an uncommon clinical entity [1]. These are present from birth, enlarge progressively with age and increase rapidly following traumatic exposure, infection and hormonal changes [1]. The patients can have varied presentations ranging from asymptomatic swelling to debilitating complications according to the vessel involved [2]. However, there are no consensus reporting guidelines and management protocols which creates a significant challenge for clinicians to manage these complex malformations successfully [2, 3]. We present a case of a 36-year old lady with a combined vascular malformation in the right side of the neck successfully managed by surgical interventions. This manuscript has been reported in line with the SCARE 2020 criteria [3].

## 2. Case report

A 36-year-old female presented to the Cardiothoracic Vascular Surgery Unit of the Department of Surgery, Dhulikhel Hospital, with a history of swelling in the right side of the neck for three months (Fig. 1). The patient's swelling progressively increased and was associated with on and off pain, especially aggravated by lying down on the right side. The patient denied the presence of swelling since early childhood, history of trauma or any record of previous surgery. However, she gave the history of using Medroxyprogesterone (Depo-Provera) discontinuously for nine years.

On clinical examination, there was a solitary oval swelling measuring 3 × 4 cm in the neck region just above the middle one-third of the clavicle. The swelling was soft, mobile, compressible, non-fluctuant, non-tender, non-pulsatile, with no overlying skin and temperature

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**Fig. 1.** Swelling on the right side of the neck measuring 3 × 4 cm.

changes. On auscultation, no bruits were heard. The cervical and axillary lymph nodes were also not palpable.

Doppler ultrasonography showed features of combined vascular malformation with venous predominance, with a velocity of 20 cm/s and an arterial component with 110 cm/s. Also, a few cystic areas which were void of flow were noted, suggestive of lymphatic malformation. Magnetic resonance imaging (MRI) was done to confirm the diagnosis. It revealed the presence of a 5 × 4 cm mass in the right side of the neck in the supraclavicular region containing lymphatic, venous, and arterial

components. **Fig. 2.**

For the management, intralesional steroid injection—40 mg Triamcinolone was administered in the perivascular area especially targeting the arterial component. One week follow-up showed a minute resolution of malformation. Following this, sclerotherapy targeting the venous and lymphatic components was done using Injection Polidocanol 2 ml which showed partial resolution of the lesion. Then, we opted for surgical excision of the lesion. Under general anaesthesia, ligation of the arterial feeders and prominent vessels was done, and lymphatic and venous components were excised. Her histopathology report suggested combined vascular malformation with lymphatic, venous and arterial components.

She was discharged on the fourth postoperative day as there were no complications at the wound site and no vascular compromise. Additionally, the patient was advised to follow some postoperative neck rehabilitative exercises as suggested by the physiotherapist. One month follow-up revealed complete resolution of the lesion with no complications.

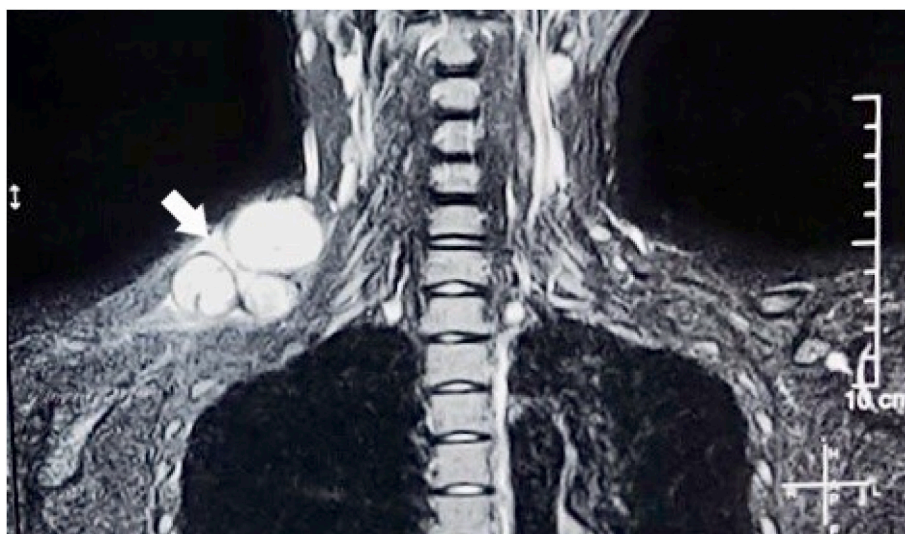
### 3. Discussion

Vascular anomalies are a group of disorders formed due to abnormal vascular development forming dysplastic vessels [2,4]. Over time, different classification systems have been introduced to properly diagnose these lesions, including Mulliken and Glowacky classification, Hamburg classification, and the most widely accepted International Society for the Study of Vascular Anomalies (ISSVA) classification [5].

Modified ISSVA 2018 classification system broadly divides vascular anomalies into two types – vascular tumours and vascular malformations [6]. Vascular malformations are further grouped into simple and combined malformation based upon the number and types of vessels involved [6]. Combined vascular malformation is defined as two or more vascular malformations (capillary, venous, arteriovenous, lymphatic) found in one lesion [6]. However, this classification system does not address a combined arterial, venous and lymphatic malformation without capillary involvement, which is the peculiarity of our case.

These malformations are presented since birth and remain obscure for an extended period [2]. Rapid enlargement of these lesions occur due to hormonal changes and traumatic incidents and do not regress independently, often leading to substantial morbidity [2]. They primarily affect the head and neck region, followed by extremities and thorax[2].

Although there is no exact etiopathogenesis, the literature suggests that most cases of vascular malformation occur sporadically with few



**Fig. 2.** T2 weighted MRI of the neck and chest coronal view showing hyperintense lesion (white arrow).

familial cases [7]. The solitary lesions are observed sporadically, whereas multiple lesions are observed in familial forms [7]. Vascular malformation has varied clinical presentation depending on the type of vessels involved [2]. These primarily include the presence of a mass, pain and functional deformity hampering the patients quality of life [2].

Due to the rarity of these lesions, there are no uniform reporting guidelines and standard management protocols. It creates an intricacy in treating these lesions, warranting a multidisciplinary approach for proper treatment [2]. Clinicians must first accurately diagnose its type, location, extension, and hemodynamic alterations [5]. Initially, detailed history taking and proper physical examinations should be taken into account [1,8], followings which, diagnostic tests like Doppler ultrasonography, X rays, CT scans, MRI, angiography, phlebography or lymphangio-scintillography can be done to confirm the type of malformation and its extent [5]. For the treatment, a multi-staged treatment with surgical intervention like embolisation of the involved vessel, sclerotherapy and intraoperative resection of the lesion is recommended [9].

Dieng et al. [10] reported a case of arteriovenous malformation (AVM) of the neck, for which they performed a two-staged treatment. Firstly, they did the ligation of the feeding vessel to reduce the arterial flux in the malformation, following which, complete ablation of the AVM was done without significant bleeding [10]. Unlike their case, our patient, had a combined malformation; hence we opted for a three-staged treatment including–intralesional steroid, sclerotherapy and surgical resection of the malformation.

Commonly, embolisation is done as the first treatment modality to reduce the risk of excessive bleeding during surgery [11]. However, as it is not affordable to the patients in our setting, we administered intralesional steroids especially targeting the arterial component, which is an affordable and safe method for involution of the malformation [12]. This mode of delivery of steroid also reduces the risk of cushingoid syndrome, and flaring of infection, which might be observed with long term systemic steroid administration [13]. Additionally, it decreases the risk of excessive intraoperative bleeding and the need for extensive surgeries in the future [12]. Several works of literature have reported the use of this modality in treating vascular anomalies [12,13].

Following intralesional steroid administration, the patient showed a slight mass reduction. Next, sclerotherapy was done targeting the venous and lymphatic components. Sclerotherapy is considered the treatment of choice either for curing the malformation or for preoperative use as it eases the surgical excision by well defining the mass and causing thrombosis of the malformed vessel [14]. Moreover, this procedure can be performed without anaesthetics and has fewer side effects [1,15].

Azizkhan et al. suggested that surgical excision of vascular anomalies not involving the vital structures and located at a convenient location yield a good outcome with a rare chance of recurrence [1]. We performed an intraoperative excision of the remaining malformation, ensuring no malformations were left. The patient is under regular follow-up, and on recent follow-up, the swelling had not recurred, and Doppler ultrasonography revealed no underlying malformation.

All and all, a rare case of combined vascular malformation must be thoroughly evaluated with detailed clinical and radiological evaluation. Furthermore, a multi-staged treatment with the administration of intralesional steroids followed by sclerotherapy, intraoperative ligation of the arterial feeders and surgical resection of the malformation should be done to ensure complete removal of the malformation.

#### 4. Conclusion

Multistage treatment modalities, including intralesional steroid injection, sclerotherapy, and surgery, are the mainstays of treating these complex vascular malformations. Regular follow-up is necessary to monitor the outcome and ensure that the malformation has not relapsed.

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#### Ethical approval

N/A.

#### Consent

Written informed consent was taken from the patient and none of the identifying characteristics were included in the original article.

#### Author contribution

First Robin Man Karmacharya Surgical procedure, patient care, manuscript writing, Guarantor, Second Satish Vaidya Surgical procedure, Patient care, manuscript writing. Third/Corresponding Swechha Bhatt Manuscript writing and editing. Fourth JN Milan Manuscript writing. Fifth Kamana Gyawali Manuscript writing Sixth Sujita Marasini Manuscript writing, Seventh Diwas Karkee Manuscript writing, Eighth Garima Gyawali Manuscript writing.

#### Registration of research studies

N/A.

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#### Declaration of competing interest

None.

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#### Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.amsu.2022.103531>.

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