

## A Case of Acquired Arteriovenous Malformation of Lip: Clinical and Mucoscopic Evaluation

A 31-year-old female presented with a slowly enlarging swollen painless lesion over left side of upper lip having occasional bleeding for 3 years with facial asymmetry. The development of lesion was preceded by a lip injury encountered in a road traffic accident 4 years back. On examination, there was presence of multiple erythematous to bluish translucent papules on mucosal side of lip over an ill-defined reddish-brown mucosa with some bleeding points [Figure 1]. On palpation, the lesions were soft, nonreducible, partially blanchable, and pulsatile with a palpable thrill. Mucoscopy was carried out using DermLite DL3 according to the method suggested by Jakhar and Grover<sup>[1]</sup> with precautions taken to prevent exertion of pressure onto the lesion to avoid any collapse of the vessels. Mucoscopy revealed bluish-red homogenous area (representing dilated arterio-venous channels in upper dermis) with nonarborizing telangiectasias (due to multiple thick-walled muscle containing blood vessels in upper and mid dermis), irregularly shaped and sized linear vessels (due to thin-walled dilated vessels), whitish structures (due to fibrotic stroma), and delicate peripheral

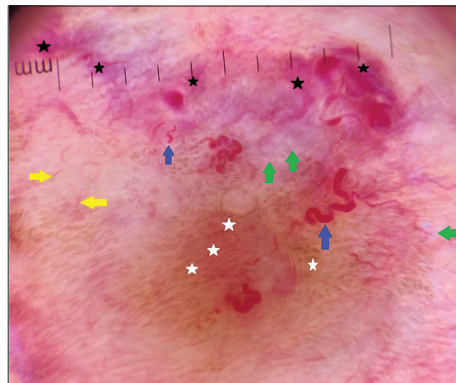


**Figure 1:** Multiple erythematous to bluish translucent papules on mucosal side of left upper lip, over an ill-defined, reddish-brown mucosa with some bleeding points

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pigment network (corresponding to basal layer hyperpigmentation and hemosiderin deposition) [Figure 2]. Once the visualization was complete, the spacer of dermatoscope was removed and cleaned with 70% isopropyl alcohol. Doppler ultrasonography indicated fast-flow high vascularity with arteriovenous shunting on spectral analysis [Figure 3], which further got confirmed by contrast-enhanced computed tomography (CECT) examination revealing left superior labial artery to be the feeding vessel. However, other routine hematological investigations and X-ray and ECG analyses ruled out any bony or cardiovascular involvement. Histopathologically, lobular arrangement of thick and thin walled vessels was present in the dermis [Figure 4]. On the basis of clinical examination and above imaging, the patient was diagnosed with an acquired



**Figure 2:** Mucoscopy revealed bluish-red homogenous area representing dilated arterio-venous channels in upper dermis (black stars), with nonarborizing telangiectasias due to multiple thick-walled muscle containing blood vessels in upper and mid dermis (blue arrow), irregularly shaped and sized linear vessels due to thin-walled dilated vessels (yellow arrow), whitish structures representing fibrotic stroma (green arrow) and delicate peripheral pigment network corresponding to basal layer hyperpigmentation and hemosiderin deposition (white stars); DermLite DL3 at 10x magnification

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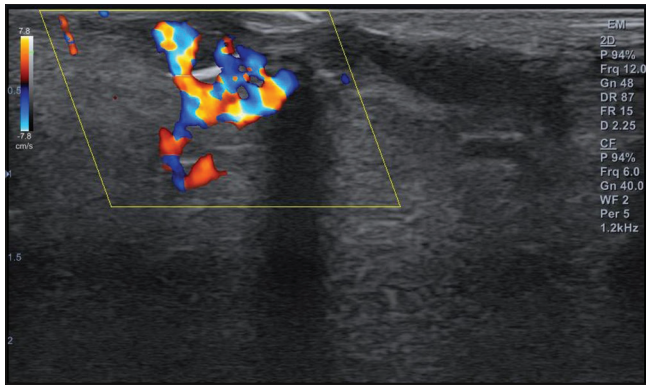
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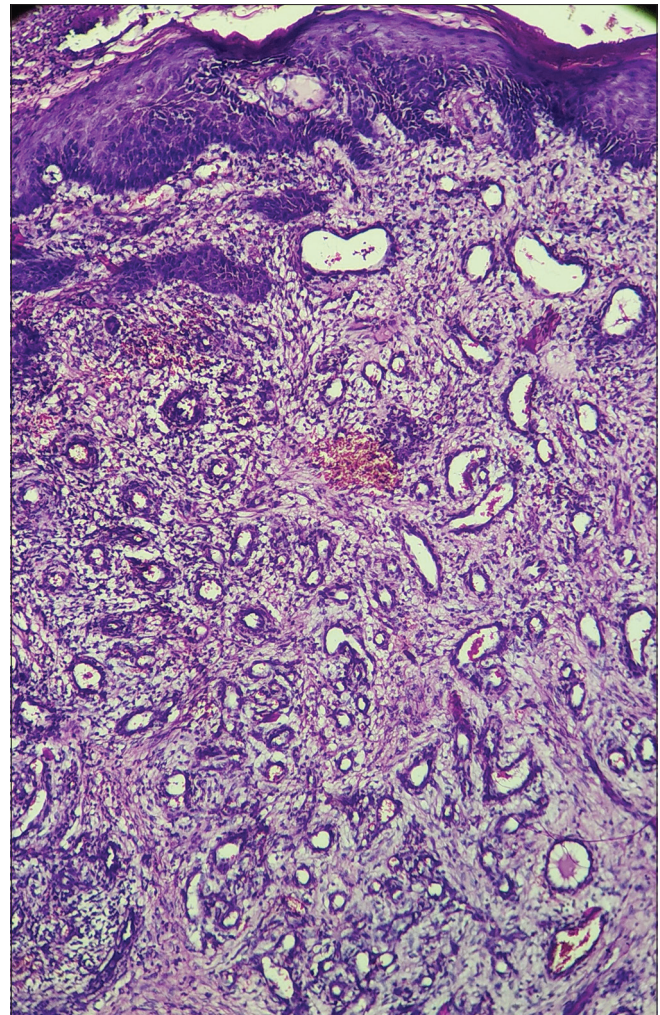


**Figure 3: Fast-flow high vascularity with arteriovenous shunting on Doppler Ultrasonography**

fast flow arterio-venous malformation (AVM) and advised a complete surgical excision with preoperative embolization, for which she denied.

AVM are channels of direct communication between arteries and veins bypassing the capillary plexus seen in congenital (defective TGF- $\beta$ ), familial (RASA1 gene mutation), and acquired forms. Generally, a history of trauma, surgery, or hormonal imbalance precedes acquired AVM leading to aberrant vascular proliferation. Unlike congenital forms (having multiple feeding vessels), acquired AVM generally have a single feeder. They progress through 4 clinical stages with local destruction and ischemia seen in stage 3 and high-output cardiac failure seen in stage 4.<sup>[2]</sup> Only 0.1% of AVM occur in head and neck with just 8.1% being extracranial. They are among the most notorious vascular lesions due to the risk of uncontrolled bleeding while performing any invasive oral procedure or examination. The most sensitive diagnostic imaging for AVM is angiography since it can delineate flow, feeding vessels, and dangerous anastomosis. However, dermoscopy is also an emerging safe and noninvasive diagnostic modality that can be used as an alternative for oral mucoscopy, in routine practice, according to the technique suggested by Jakhar and Grover.<sup>[1]</sup> This technique also aids in avoiding any direct contact and contamination of a dermatoscope.

Dermoscopically the absence of lacunae is the hallmark of AVM, which helps in ruling out differential diagnoses. Lacunae are clustered, well defined round to oval structures seen as a distinguishing feature of various vascular tumors. The most peculiar finding in hemangioma is red lacunae, whereas angiokeratoma characteristically shows dark and deeply pigmented lacunae. Similarly, the presence of two-toned lacunae (yellow and red) is specific for lymphangioma circumscripta. Histologically, these lacunae represent thin-walled dilated vessels in papillary dermis, whereas the prototypical thick-walled muscular vessels of AVM are seen as nonarborizing vessels on dermoscopy. The other differential diagnoses considered in our patient were pyogenic granuloma and basal cell carcinoma.



**Figure 4: Lobular arrangement of thick and thin walled vessels in dermis with large vessels surrounded by proliferating small vessels (H and E; 100x)**

Pyogenic granuloma can be ruled out dermoscopically by the presence of white rail lines over a red homogenous area. Similarly, basal cell carcinoma can be ruled out by the presence of arborizing vessels (in-focus telangiectasia with tree like ramifications), which is the most common finding in it.<sup>[3]</sup>

#### ***Declaration of patient consent***

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

#### ***Conflicts of interest***

There are no conflicts of interest.

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