

Great Radiologic Imitators: Arteriovenous Malformation of Mandible – A Case Series

Abstract

An arteriovenous malformation (AVM) is a vascular malformation characterized by anomalous communications between arteries and veins without the normal intervening capillary bed. AVMs of jaw are extremely rare conditions that can give rise to dreadful complications if handled carelessly. Fifty percent of all intraosseous AVMs occur in the maxillofacial region and are extremely infrequent in the mandible. Only six cases of extraosseous submandibular AVMs have been mentioned in the literature. Three cases of AVMs involving the mandible, with a massive case involving both the mandible and submandibular region is reported. To the best of our knowledge, only one case is documented as a combined AVM involving both the mandible and submandibular region in literature.

Keywords: Arteriovenous malformation, embolization, mandible, submandibular region

Introduction

Before the 1980s, vascular lesions were referred to as hemangiomas.^[1] In 1982, Mulliken and Glowacki suggested a binary classification system for vascular anomalies based on pathologic features.^[2] This system, which was adopted by the International Society for the Study of Vascular Anomalies (ISSVA), has since been expanded and is nowadays extensively accepted.^[3] The ISSVA classification system splits vascular anomalies into two primary biological categories: (1) vasoproliferative or vascular neoplasms (example: infantile hemangioma, congenital hemangioma, etc.) and (2) vascular malformations (for example: arteriovenous malformation [AVM], arteriovenous [AV] fistula, etc.).^[2]

The two classes of vascular lesions have basically different etiologies and clinical presentations. Hemangiomas exhibit endothelial hyperplasia and expand by cellular proliferation. Clinically, hemangiomas usually appear in early infancy, grow rapidly during the 1st month of life, then gradually involuting over 5 or 6 years.^[3,4] On the other hand, AVMs do not exhibit endothelial proliferation, instead display progressive ectasia of abnormal vessels, lined by flat endothelium. AVMs

occur as a result of errors of vascular morphogenesis that is always present at birth, grow proportionally with the child, and may evident at any time during life due to an events such as trauma, surgery, and infection.^[4,5]

Vascular malformations can be subdivided based on the rate of blood flow: “slow flow” (capillary malformation, venous malformation, or lymphatic malformation), “fast flow” (arterial malformation, AVM, or AV fistulae), and combined vascular malformations.^[6] AVMs are characterized by a collection of arterial and venous channels without a significant solid recognizable mass.^[7] Histologically, AVMs consist of dysplastic arteries that drain into arterialized veins making a vascular nidus bypassing capillary beds. The prevalence of AVMs is unknown, but estimates range from 5 to 613/100,000 persons.^[8] AVMs may present with pain, ulceration, ischemic changes, bleeding, and congestive heart failure. On physical examination, they may be warm pink patches on the skin with an underlying vascular murmur or thrill. Common locations include intracranial, intraosseous, muscle, and subcutaneous fat.^[6] The management and the therapy of this condition in the maxillofacial region are difficult because of the abundant vascular network.^[5,9]

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**A. K. Nabeel,
Jesmith Elsa Jacob,
Tinky Bose,
K. P. Sangeetha,
K. N. Sandhya,
Anita Balan**

*Department of Oral
Medicine and Radiology,
Government Dental College,
Thiruvananthapuram, Kerala,
India*

Address for correspondence:

*Dr. A. K. Nabeel,
Government Dental
College, Room No. 203
Medical College P O,
Thiruvananthapuram - 695 011,
Kerala, India.
E-mail: nabeelsabeel@gmail.
com*

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Case Reports

Case 1

A 29-year-old male patient reported with a complaint of swelling of the right lower face for the past 8 years. The patient gave the history of extraction of lower right molar tooth 8 years back, after which he noticed a small swelling. This progressively increased to the present size. No pain was associated with it. The patient also had paresthesia of the right side of the lower lip of 1-year duration. Family and medical history were noncontributory.

Facial asymmetry was observed owing to a swelling on the right side of the mandible. Clinical examination revealed diffuse swelling on the body, the angle, and the ramus of mandible on the right side. The swelling extended anteroposteriorly from the midline to posterior border of the ramus of mandible, and superoinferiorly from 2 cm below the ala-tragus line to 2 cm crossing the inferior border of the mandible [Figure 1]. No visible pulsations were seen. The swelling was nontender and soft in consistency and compressible with no local rise in temperature. Palpable thrill was also noted.

On intraoral examination, approximately a 10 cm × 4 cm size swelling was noticed on the right alveolus involving the retromolar trigone and the floor of mouth which crossed the midline. Right lower posterior teeth were pushed medially. Right tonsil was also shifted medially and the tongue was pushed posteriorly [Figure 2].

A provisional diagnosis of vascular malformation was given based on palpable thrill and history.

Occlusal radiograph showed buccal expansion with altered trabeculae and the absence of cortical outline on the right side of the mandible. Medial drifting of right posterior teeth was noted [Figure 3]. Panoramic radiograph showed a mixed lesion with predominantly radiopaque areas, increased marrow spaces, and increased height of the right body of the mandible. Widening of right inferior alveolar canal and thinning of right inferior cortical border of the mandible was noted [Figure 4].

Ultrasonography of face and neck was done using 12-MHz transducer which revealed the increased vascularity in color flow Doppler imaging over the right body of the mandible compared to the contralateral side [Figure 5].

Contrast-enhanced computed tomography (CT) of mandible revealed enlargement with heterogeneous density in the body, ramus, and angle of the mandible on the right side. Multiple dilated vascular channels were seen on the right lower alveolus, right floor of mouth, right submandibular space, and right sublingual space. Multiple dilated veins were draining into the right anterior jugular vein and the right retromandibular vein. Communication between right anterior jugular vein and right internal jugular vein was also noticed. The right anterior jugular vein was dilated.

The lesion was mainly fed by the right inferior alveolar artery, which was dilated and tortuous [Figure 6].



Figure 1: Extraoral view showing facial asymmetry owing to the swelling on right body of mandible (Case 1)



Figure 2: Intraoral view showing swelling on right alveolus involving the retromolar trigone and the floor of mouth which crosses the midline (Case 1)

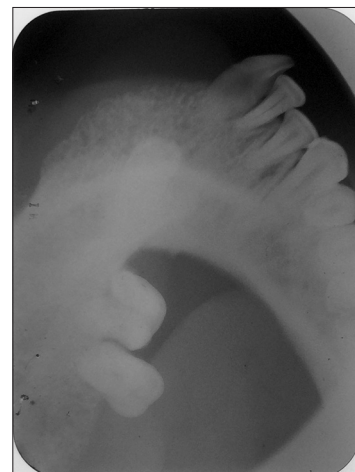


Figure 3: Intraoral occlusal radiograph showing buccal cortical expansion with altered trabeculae and absence of cortical outline on the right side of the mandible (Case 1)

Under local anesthesia through right femoral arterial approach, right external carotid angiogram was done, which revealed evidence of AVM in the right mandible with feeding arteries from inferior alveolar (branch of internal maxillary artery) and facial artery both of which were dilated and tortuous and were draining into jugular vein [Figure 7]. The diagnosis of a massive AVM on the right side of mandible was confirmed.

Wide resection of the affected part of the mandible after embolizing the feeder vessels under general anesthesia and reconstruction with titanium plates was planned. However, the patient refused the treatment. He is under observation.

Case 2

An 11-year-old female patient reported with a complaint of swelling on the left side of the lower face for 2 weeks. The patient gave the history of extraction of lower left molar tooth 3 weeks back, following that she had severe bleeding from the extraction socket which resulted in hypovolemic shock. The bleeding was initially controlled by packing the extraction socket and blood transfusions were required. After 1 week, she noticed a small swelling on the same site, which gradually increased in size and reached the present size. No pain was associated with it. Family and medical history were noncontributory.

On extraoral examination, facial asymmetry was observed owing to a swelling on the left side of the mandible. Clinical



Figure 4: Panoramic radiograph showing mixed lesion with predominantly radiopaque area, with trabecular alteration, increased marrow spaces, increased height of right body of mandible, and widened right inferior alveolar canal (Case 1)

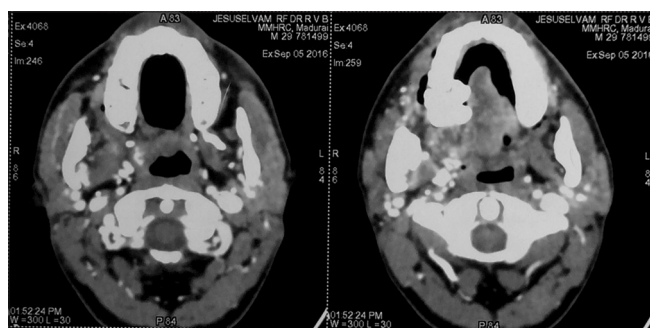


Figure 6: Contrast-enhanced computed tomography revealed enlargement with heterogeneous density in the body, ramus, and the angle of right mandible (Case 1)

examination revealed diffuse swelling of the body of the mandible. The swelling extended anteroposteriorly from the angle of mouth to anterior border of the ramus of mandible on the left side and superoinferiorly from at the level of angle of mouth to 1 cm crossing the inferior border of the mandible [Figure 8]. Surface texture of the swelling was normal and no visible pulsations were seen. The swelling was nontender and firm in consistency; compressible, palpable thrill was also noted. There was no local rise in temperature.

On intraoral examination, mild reddish enlargement of buccal gingiva in relation to 36 was noted. Buccal cortical expansion was noted in relation to 34, 35, and 36. The swelling was nontender and firm in consistency; compressible, palpable thrill was also noted [Figure 9].

A provisional diagnosis of vascular malformation was given based on palpable thrill and history.

Panoramic radiograph revealed altered trabecular pattern with multiple radiolucent spaces on left body of mandible,

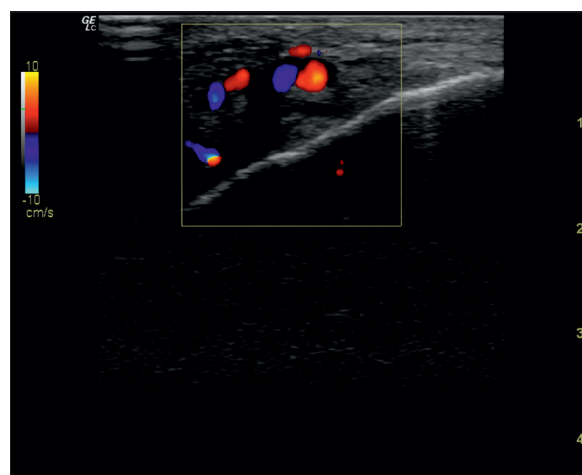


Figure 5: Ultrasonography revealing the increased vascularity on right body of mandible region (Case 1)

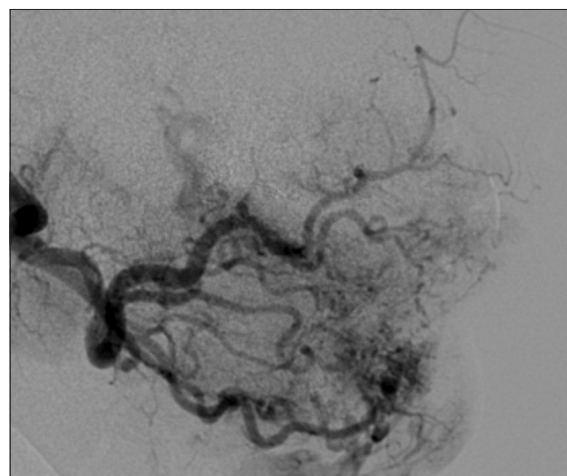


Figure 7: Right external carotid angiogram showing feeding arteries from inferior alveolar artery and facial artery, both of which are dilated and tortuous, and are draining into jugular vein (Case 1)

increased vertical height of left body of the mandible, and dilated inferior alveolar canal on the left side, which could not be traced anteriorly. Periodontal space widening and loss of lamina dura noticed in relation to 34, 35, and 36 [Figure 10].

CT of mandible revealed a lesion of regular expansive soft-tissue density on the left angle of mandible, with cortical erosion and sclerotic margins. No cystic areas or calcification was noticed. Inferior alveolar canal appeared dilated with mild soft-tissue density within [Figure 11].

Magnetic resonant imaging (MRI) with contrast was done. The left half of mandible was expanded with bony changes. An irregular cavity was noted in the posterior part where mixed signal tissue was seen. The entire left inferior alveolar canal was expanded. Multiphase angiogram showed tortuous vascular channels in the premolar region, mainly fed by the inferior alveolar artery, which drains into external jugular vein [Figure 12]. This confirms the diagnosis of an AVM on the left side of mandible.

Under general anesthesia by nasoendotracheal intubation, after embolizing the feeder vessels, the mandible was resected *en bloc* with the pathologic contiguous soft tissue

from the area of the left ramus to the distal aspect of left canine. After resection and removal of the involved mandibular segment and control of hemorrhage, teeth 34, 35, and 36 were extracted from the segment, and the convoluted vascular mass was curetted. The remaining cortical shell was then grafted with a corticocancellous block of bone from the right fibula. The patient is under regular follow-up.

Case 3

A 15-year-old female patient reported with a complaint of swelling on the left side of the lower face for 3 years. She noticed progressive increase in size of the swelling and reached the present size. She also noticed increasing asymmetry of the face. No pain was associated with it. Family and medical history were noncontributory.

On extraoral examination, facial asymmetry was observed owing to an hyperpigmented swelling on the left side of the mandible. Clinical examination revealed soft, nontender, hyperpigmented swelling lateral to left chin



Figure 8: Extraoral view showing facial symmetry owing to swelling on the left side of the mandible (Case 2)



Figure 10: Panoramic radiograph showing altered trabecular pattern with multiple radiolucent spaces on left body of mandible, increased vertical height of left body of mandible, and dilated left inferior alveolar canal on left side, which cannot be traced anteriorly (Case 2)



Figure 9: Intraoral view showing mild reddish enlargement of buccal gingiva in relation to 36 (Case 2)



Figure 11: Computed tomography scan showing regular expansive soft-tissue density lesion on left angle of mandible, with cortical erosion and sclerotic margins (Case 2)

region 2 cm below the corner of mouth and 1 cm above the lower body of mandible [Figure 13]. On inspection, no visible pulsations were noticed. On palpation, the swelling was nontender and soft in consistency and compressible. Pulsation and bruit were noticed just below the left corner of mouth and submandibular region. There was no local rise in temperature.

No visible swelling was noted intraorally. There was mobility of 34, 35, 36, and 37 and 35 and 36 depressed on applying pressure. Pulsation and bruit in relation to buccal vestibule of 35 and 36 were noticed [Figure 14].

A provisional diagnosis of vascular malformation was given based on palpable pulsation, bruit, and history.

Occlusal radiograph showed mild expansion of buccal cortical plate in relation to 36, 37, and 38 regions [Figure 15]. Panoramic radiograph revealed ill-defined radiolucent lesion with poorly defined margins confined to the left body of mandible extending from distal aspect of 32 till radicular aspect of developing tooth germ 38. Root resorption of 34, 35, 36, and 37 with loss of

lamina dura noticed. Altered trabecular pattern, thinning of inferior cortical border of right mandible, and dilated inferior alveolar canal on left side, which cannot be traced anteriorly was also noticed [Figure 16].

Ultrasonography of face and neck was done using 12-MHz transducer which revealed the increased vascularity in the course of left facial artery anterior to left masseter in color flow Doppler imaging [Figure 17a]. Increased vascularity also was noticed over the vessels of left submandibular gland [Figure 17b].

Contrast-enhanced CT of mandible revealed large expansile lesion on left body of the mandible with brilliant enhancement after intravenous contrast and minimal rarefaction of lingual and labial cortex with very rich vascularity. Prominent serpiginous nidus of vessels seen at left half of mandible and adjacent subcutaneous compartment supplied through branches of external carotid arteries such as lingual, facial, and superficial temporal arteries representing AVM of the left mandible [Figure 18].

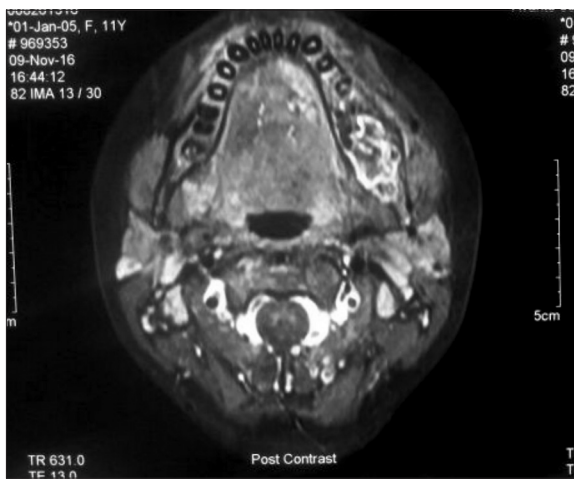


Figure 12: Magnetic resonance imaging with contrast showing left half of mandible expanded with bony changes (Case 2)



Figure 13: Extraoral view showing hyperpigmented swelling lateral to the left chin, 1 cm above the inferior border of mandible (Case 3)



Figure 14: Intraoral view showing no visible swelling (Case 3)

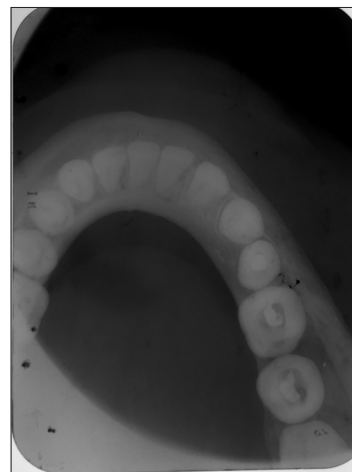


Figure 15: Intraoral occlusal radiograph showing mild expansion of buccal cortical plate on left body of mandible in relation to 36, 37, and 38 regions (Case 3)

Under local anesthesia through left femoral arterial approach, left external carotid angiogram was done, which revealed evidence of AVM in the left side mandible with feeding arteries from branches of left external carotid arteries such as lingual, facial, and superficial temporal arteries which were dilated and tortuous [Figure 19]. This confirmed the diagnosis of AVM on the left side of mandible.

Under general anesthesia by nasoendotracheal intubation, after embolizing the feeder vessels, the mandible was resected *en bloc* with the pathologic contiguous soft tissue from the area of the left ramus to the mesial aspect of the left lateral incisor. After resection and removal of the involved mandibular segment and control of hemorrhage, teeth 32, 33, 34, 35, 36, 37, and 38 were extracted from the segment and the convoluted vascular mass was curetted. The remaining cortical shell was then grafted with a corticocancellous block of bone from the right fibula. The patient is under regular follow-up till now.

Discussion

AVMs are extremely rare lesions that can be life-threatening if left untreated due to enormous blood loss during tooth extraction or biopsy. Although the head and neck constitute <14% of the total surface area of the body, approximately 50% of all vascular malformations occur in this region but are extremely infrequent in the mandible.^[10]



Figure 16: Panoramic radiograph showing ill-defined radiolucent lesion with poorly defined margins confined to left body of mandible extending from distal aspect of 32 till radicular aspect of developing tooth germ 38. Root resorption of 34, 35, 36, and 37 with loss of lamina dura also noticed (Case 3)

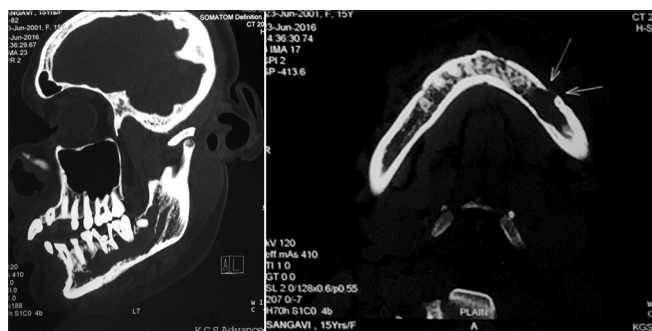


Figure 18: Contrast-enhanced computed tomography showing large expansile lesion on left body of mandible with brilliant enhancement and minimal rarefaction of lingual and labial cortex with very rich vascularity (Case 3)

Intraosseous AVMs of the jaws are relatively rare, with fewer than 200 cases reported in the literature.^[11] Only 6 cases of extraosseous submandibular AVMs have been mentioned in the literature (5 females and 1 male). To the best of our knowledge, the male patient reported here will probably be the second documented case of combined AVM involving both the mandible and submandibular region.

AVMs usually appear in adolescence but have an age range of 3 months to 74 years. Some authors noted predominance in females (female-to-male ratio, 2:1) while others have reported equal prevalence among males and females.^[1] AVMs usually present with nonspecific symptoms including bruit, dental loosening, swelling of soft tissues, change in skin and mucosal color, and dysesthesia of the lower lip or chin.^[12]

Radiographically, vascular malformations of the jaws have been referred to as the “great radiologic imitators” and can look like any lesion ranging from a cyst to a malignancy.^[13] In the mandible, vascular malformation produces an ill-defined, radiolucent image, often with the appearance of honeycomb or soap bubbles, with small, rounded, and uneven lacunae or a punched-out area. AVMs most commonly appear as multilocular radiolucencies in panoramic examinations with dilated inferior alveolar canal and widening of the marrow spaces.^[14]

Thus, a large number of tumors, both benign and malignant, should be considered in the differential diagnosis. Of the benign lesions, squamous cysts are the most frequent. Less common lesions include nonepithelial cysts (also known as hemorrhagic or traumatic bone cysts), fibrous dysplasia, fibroma, myxoma, neurofibroma, eosinophilic granuloma,

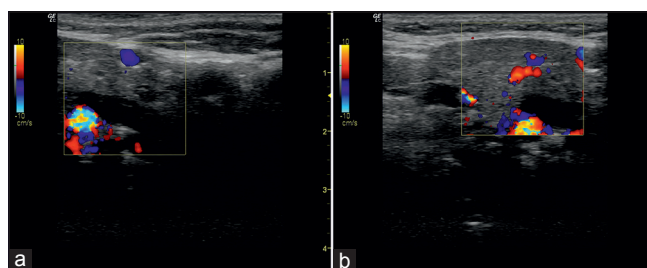


Figure 17: (a) Ultrasonographic image showing the increased vascularity in the course of left facial artery anterior to left masseter in color flow Doppler imaging (Case 3). (b) Ultrasonographic image showing the increased vascularity over the vessels of the left submandibular gland (Case 3)

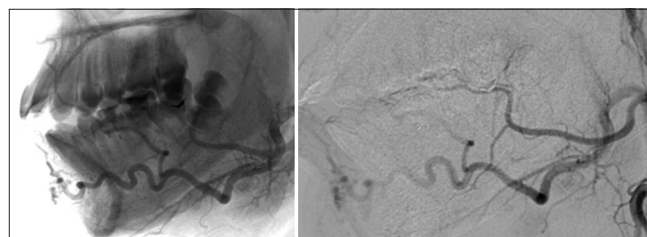


Figure 19: Left external carotid angiogram showing arteriovenous malformation on the left side of mandible with feeding arteries from branches of left external carotid arteries such as lingual, facial, and superficial temporal arteries which were dilated and tortuous (Case 3)

and aneurysmal bone cysts.^[15] Since it usually appears as a multilocular radiolucency, other radiographic aids such as CT, MRI, and digital subtraction angiography (DSA) are thought to gain a precise diagnosis of AVM.

Management of AVMs is usually complex and necessitates a multidisciplinary team for fruitful outcome. Observation may be used as a transitory measure in special situations, such as extreme age, pregnancy, or refusal of therapy.^[12] Arterial ligation was used in the past as a purely symptomatic treatment or before surgery. At present, it is well known that ligation of external carotid artery should not be done, firstly as many anastomoses encourage the rapid appearance of a collateral circulation, and secondly because future embolization would be impossible.^[16]

At present, superselective angiographic embolization is considered as first-line treatment, alone or in combination with surgical method to reduce intraoperative bleeding. Occlusion of the lesion is gained using movable balloon, coils, or liquid glue.^[17] However, serious complications after embolization (e.g., occlusion of pulmonary or cerebral vessels) or recurrence of AVMs should be considered. Intraosseous injection of sclerosing agents should be used to achieve further obliteration of the AVMs.^[18] For treating the AVMs of the mandible, block resection of the affected area has been suggested and temporary reconstruction with alloplastic bone plate or with the patient's own free, previously curretted mandibular segment has been reported.^[19]

Conclusion

AVMs have no characteristic feature on plain film radiography. Although AVM is rare, the operator must always consider it and should add image inspection with new imaging modalities such as CT, MRI, and DSA and intraosseous AVMs should always be included in the differential diagnosis of multilocular radiolucent jaw lesions. Dentists must be aware of the clinical manifestations of AVM, to prevent iatrogenically related accidents and to minimize potential spontaneous crisis to the patient.

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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Conflicts of interest

There are no conflicts of interest.

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