International Journal of Neurosurgery
2025, Vol. 9, No. 1, pp. 10-16
https://doi.org/10.11648/j.ijn.20250901.12

Case Report

A Rare Case of External Ear Arteriovenous Malformation Treated with Linac Based Radiosurgery

Mariya Deputy*, Sanjay Hunugundmath, Amit Nirhali, Vishram Naik, Sharad Gadhave

Department of Radiation Oncology, Sahyadri Super Speciality Hospitals, Pune, India

Abstract

Background: Arteriovenous malformation (AVM) is a congenital vascular anomaly in which there are abnormal connections between arteries and veins in the form of fistula or nidus without an intervening capillary bed. External ear AVMs can cause swelling, redness, bleeding, and pain. Timely diagnosis and treatment are crucial to prevent complications. According to literature, there are different techniques to treat auricular AVM which include surgical excision followed by ear reconstruction, sclerotherapy and embolization. Stereotactic radiosurgery (SRS) has an established role in treating intracranial AVM with excellent obliteration rates but limited literature exists on its efficacy in treating auricular AVM in terms of obliteration rates and cosmetic outcome. *Case Presentation*: We report a rare case of right external ear AVM which was post embolization and post excision followed by reconstruction but had persistent residual nidus in right pinna treated by stereotactic radiosurgery. He was treated to a dose of 21 Gy in 3 fractions at 7 Gy per fraction for 3 consecutive days. He had completed treatment without any complications. Follow up after 2 weeks of treatment, clinically, patient had redness, swelling and persistent pulsations but no episode of bleeding. At the six-month follow-up, redness, swelling, and pulsations had decreased. Follow up after 1 year post treatment, clinically, redness, swelling and pulsations were not present. Dynamic Brain MRI with angiography showed complete obliteration of residual AVM nidus in right pinna. Patient was satisfied with the cosmetic outcome. *Conclusion*: We conclude that stereotactic radiosurgery can be used as an effective treatment modality for auricular AVMs. In our case, radiosurgery provided an excellent control and obliteration of the nidus and good cosmetic result eliminating the need for surgical reconstruction at the site of the nidus.

Keywords

Arteriovenous Malformation, Digital Substraction Angiography, External Ear, Nidus, Dynamic Contrast Enhanced Mri, Onyx, Embolization, Excision

1. Introduction

Arteriovenous malformation (AVM) is a congenital vascular anomaly characterized by abnormal connections between arteries and veins, forming fistulas or nidus without an intervening capillary bed. These high-flow lesions cause oxygenated blood to bypass the capillaries and flow directly into the venous system, leading to ischemia, bleeding, pain, and ulceration [1]. The head and neck region is the most common site of peripheral AVM, with auricular AVMs being

*Corresponding author: deputymariya371@gmail.com (Mariya Deputy)

Received: 30 January 2025; **Accepted:** 14 February 2025; **Published:** 24 February 2025

the second most prevalent in this area [2]. Although auricular AVMs are non-malignant, they significantly impact a patient's appearance, life, and mental well-being [1]. The main goals of treating auricular AVMs are to alleviate symptoms such as pain, bleeding, and ulceration while also achieving a satisfactory cosmetic outcome. Various treatment modalities are available, including surgical excision followed by ear reconstruction, sclerotherapy, and embolization. Stereotactic radiosurgery is well-established as an effective treatment for intracranial AVMs, with excellent obliteration rates, but its efficacy in treating auricular AVMs remains poorly documented. To address this gap in the literature, we present a rare case of an external ear AVM treated with stereotactic radiosurgery. To the best of our knowledge, this is the first documented instance of such a treatment for auricular AVMs.

2. Case Presentation

A 29 year old male with no history of comorbidities, significant personal and family history presented in April 2022 with complaints of right ear swelling, redness, bleeding episodes and increased pulsations. Digital subtraction angiography (DSA) conducted in April 2022 revealed a high-flow lesion involving the right external ear. External carotid artery (ECA) injection confirmed the presence of an enlarged, dilated, tortuous posterior auricular artery with multiple feeders leading to a large Arteriovenous malformation. According to clinical findings, it was Schobinger clinical stage III right auricular AVM (Table 1) [3].

Table 1. *Schobinger clinical staging system for peripheral AVM [3].*

Schobinger stage	Features
I (quiescence)	Cutaneous blush or warmth
II (expansion)	Bruit, audible pulsation, and enlarging lesion
III (destruction)	Pain, ulceration, bleeding, and infection
IV (decompensation)	Cardiac failure

Following embolization, the condition partially improved. However, in December 2022, the patient presented again with redness, bleeding, and increased pulsation in the right external ear. DSA performed in December 2022 showed an increase in the size of the AVM. Consequently, embolization was repeated on December 29, 2022. The final right ECA injection demonstrated near 60-70% obliteration of the AVM. Multiple minor feeders supplying the pinna AVM were also noted. On December 29, 2022, the patient underwent wide local excision of the lesion and reconstruction. Since March 2023, he had presented with a mild increase in right ear pinna swelling and increased pulsations. DSA performed on May 25, 2023, revealed an enlarged, dilated, tortuous posterior auricular artery with multiple feeders leading to a moderate sized AVM. The findings suggested the presence of a residual AVM in the right pinna. The patient was observed, and on July 4, 2023, he experienced a bleeding episode from the right external ear AVM. Repeat DSA performed on July 4, 2023, showed similar findings to those observed in May 2023. Given the patient's unwillingness to undergo re-excision or amputation of the ear, the decision was made to treat the residual nidus with stereotactic radiosurgery. Clinical photograph of patient is shown in (Figure 1).

Figure 1. *Pretreatment clinical photograph of patient. Anterior aspect (a) and posterior aspect (b) pointed with arrows.*

A Linac-based stereotactic radiosurgery system, specifically ELEKTA synergy, was planned for the treatment. The planned dose was 21 Gy in three fractions, with 7 Gy per fraction over three consecutive days. A custom-made immobilization device, consisting of a head rest and head and neck orfit, was used to ensure proper positioning of the patient. A Dynamic Brain MRI with contrast and angiography was conducted (Figures 2, 3) with a 1 mm slice thickness.

Figure 2. *Pretreatment Dynamic Brain MRI with contrast. Residual nidus is pointed with arrow.*

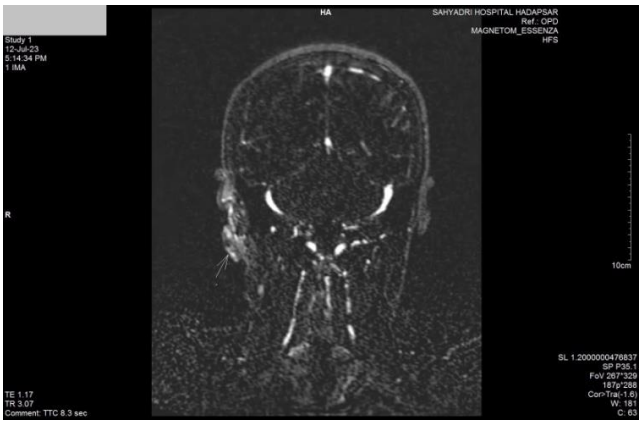


Figure 3. *Pretreatment MR Angiography. Residual nidus is pointed with arrow.*

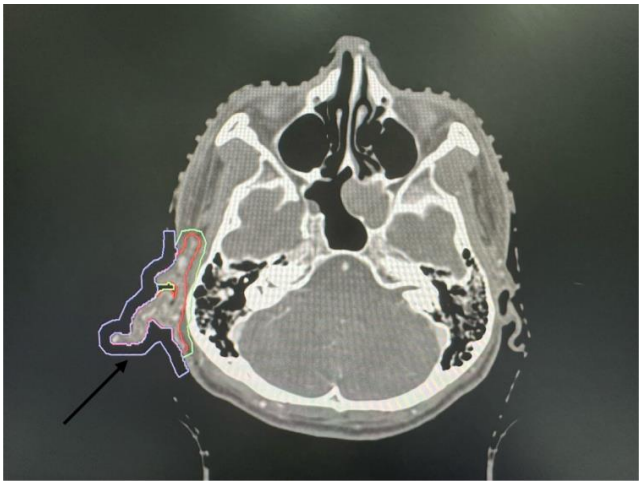


Figure 4. *Planning CT scan with contrast. It shows red, gross tumor volume and green, planning target volumepointed with arrow.*

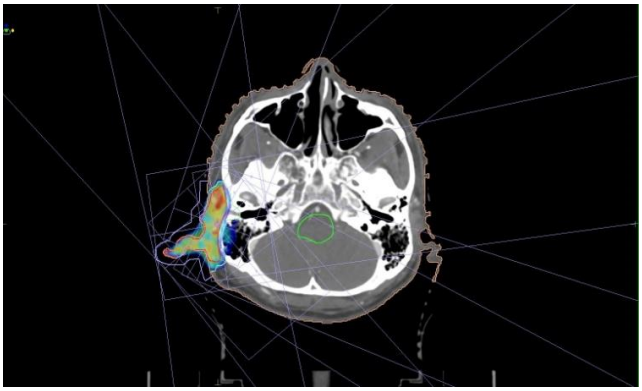


Figure 5. *IMRT plan on planning CT scan. It shows 9 field IMRT plan, virtual bolus and 100 % dose wash.*

Additionally, a planning CT brain plain with the same slice thickness was acquired. These images were imported into the Monaco planning system. T1 post-contrast MRI brain images and CT images were fused. The gross tumor volume (GTV), which encompassed the residual nidus involving the right pinna, was contoured. Subsequently, a direct 2 mm auto margin from the GTV to the planning target volume (PTV) was generated to compensate for any setup errors (Figure 4). The plan was executed using 9 field IMRT with a 06 MV Photon beam energy and virtual bolus (Figure 5). The plan was approved with 98 % V100 coverage (volume receiving 100% ofprescription dose) of GTV and 95 % V100 coverage of PTV. Target coverage and dose constraints achieved for OAR's (Organs at risk) are shown in (Table 2). Pretreatment patient specific QA (quality assurance) was conducted, and the results were satisfactory, with a 95% gamma value (Figure 6).

Table 2. *Target coverage and dose constraints of Organs at risk. OAR, Organs at risk; GTV, gross tumor volume; PTV, planning target volume; V100, volume receiving 100% of the prescription dose; Dmax, maximum point dose; Dmean, mean dose.*

Structures	Target coverage and OAR constraints	
GTV (residual nidus)	V_{100}	98 %
	Dmax	107%
PTV	V_{100}	95 %
	Dmax	107%
BRAINSTEM	Dmax	6.38 Gy
RIGHT COCHLEA	Dmean	10.73 GY
Mandible	Dmax	20 Gy

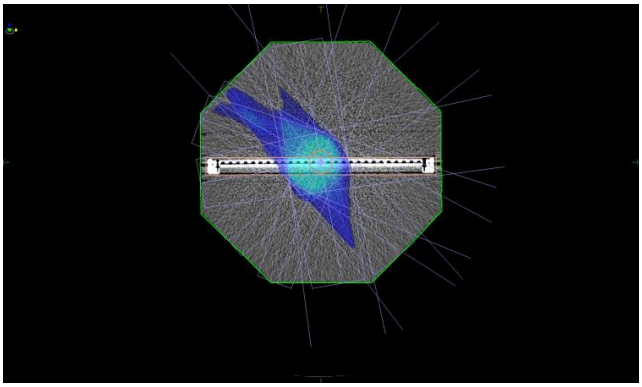


Figure 6. *Pretreatment patient specific quality assurance done with PTW OCTAVIOUS 1500 Phantom on which gamma analysis was done.*

For treatment delivery, the patient was positioned on the couch with a head rest and five clamp head and neck orfit (Figure 7). CBCT (cone beam CT scan) was taken, and after correcting any setup errors, a bolus of 5 mm was placed over the right Pinna. The patient completed the treatment in three days without experiencing any significant complications. Two weeks post-treatment, there was persistent redness, swelling and pulsations in the right external ear, but there were no

bleeding episodes (Figure 8). 6 months post treatment clinically, there was reduction in the redness, swelling and pulsations in right external ear with no episode of bleeding. On follow-up after one year of treatment, there was a notable reduction in redness and swelling, no episodes of bleeding, and pulsations (Figure 9). The patient expressed satisfaction with his appearance and cosmetic results. Dynamic Brain MRI with angiography after one year revealed complete obliteration of the residual nidus (Figures 10, 11).

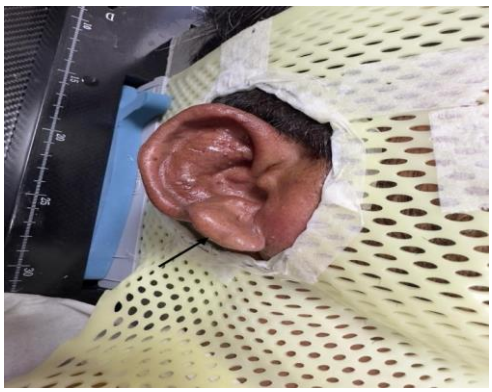


Figure 7. *Immobilization of patient on treatment couch with head and neck orfit and head rest pointed with arrow.*

Figure 8. *Clinical photograph of patient 2 weeks post treatment.*

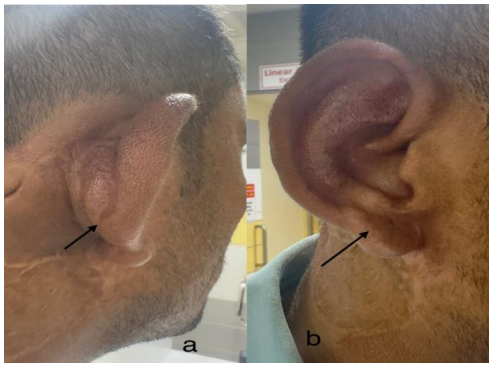


Figure 9. *1 year post treatment clinical photograph. Posterior aspect (a) and anterior aspect (b) pointed with arrows.*

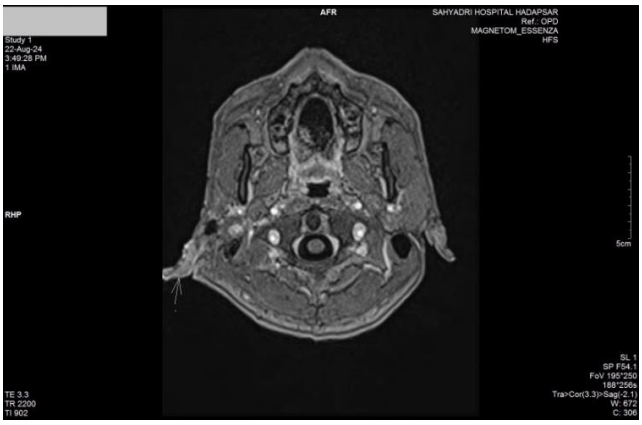


Figure 10. *1 year post treatment Dynamic Brain MRI with contrast shows complete resolution of residual nidus pointed with arrow.*

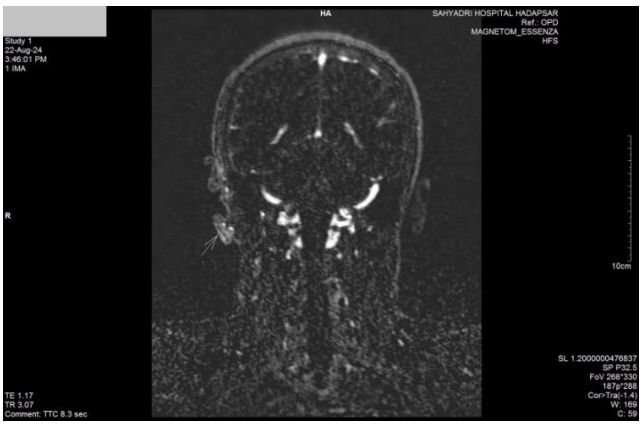


Figure 11. *1 year post treatment MR Angiography shows complete resolution of residual nidus pointed with arrow.*

3. Discussion

Arteriovenous malformation (AVM) is high blood flow vascular anomaly characterized by abnormal connections between arteries and veins. The exact cause of AVM in the ear is still unknown, but they are believed to be present at birth and may be triggered by trauma, infection, iatrogenic factors, or hormonal changes, such as puberty, pregnancy, or hormone therapy [4-12]. Due to the shunting of high-pressure blood flow from arteries directly into veins, it causes various symptoms, including warmth, swelling, redness, pulsation, fremitus, and ear noise. Furthermore, it can lead to ischemic changes, resulting in pain, ulceration, itching, and bleeding [13, 14]. The abnormal tissue surrounding cartilage can cause an overgrowth of cartilage, leading to a condition called macrotia. Typically, auricular AVMs have one to three feeding arteries, with the primary feeders being the ipsilateral superficial temporal artery, the posterior auricular artery, and the occipital artery [10, 11, 14, 15]. The treatment modalities usually used to treat auricular AVM are surgical excision +/- reconstruction, sclerotherapy, embolization or amputation. The primary goal of treatment is to obliterate nidus to prevent

bleeding as well as to achieve good cosmesis.

In 2005, Wu et al [8], studied 41 patients with auricular AVM in which progression was assessed in observation, ligation, embolization alone and amputation groups. The study showed that, there was no improvement in observation and ligation group. 39 % of patients had controlled findings and 3 patients had improvement. In 2009, Whitty et al [16], published a case report on treatment of external ear AVM in a 15 year old patient in which excision was done and there was no evidence of recurrence. In 2016, Kim et al [17], published study on combination of Embolization followed by excision in 60 year old patient with AVM of ear in which they observed that embolization followed by excision results in better removal of nidus without bleeding. In 2018, Gupta et al [18], published a case report in which preoperative coil embolization was done followed by reconstruction in 24 year old male with huge AVM. We couldn't find literature in which stereotactic radiosurgery was used as one of the treatment modality.

In our case, the patient had a Schobinger clinical stage III auricular AVM. Initially, he underwent embolization, but it resulted in recurrence, necessitating a combination of embolization, surgical excision and reconstruction. However, the residual nidus in the right pinna caused bleeding episodes. Additionally, macrotia, redness, and increased pulsations were other symptoms that caused discomfort to the patient. Given these symptoms, we decided to treat the residual nidus with stereotactic radiosurgery. The treatment involved delivering a dose of 21 Gy in three fractions, with 7 Gy per fraction, over three consecutive days. The patient successfully completed the treatment without experiencing any major complications. Two weeks after treatment, the patient showed persistent redness, swelling, and pulsations, but there were no bleeding episodes. Six months later, the redness, swelling, and pulsations had significantly reduced, and there were no bleeding episodes. A year later, Dynamic Brain MRI scan revealed complete obliteration of the AVM nidus. Clinically, the patient experienced no redness, swelling, pulsations, or bleeding episodes. He was satisfied with the cosmetic outcome. A multidisciplinary approach is crucial in diagnosing and treating auricular AVMs.

While surgery, followed by reconstruction and embolization, are the most commonly preferred treatment modalities based on literature, our case demonstrates the potential of stereotactic radiosurgery (SRS) as an effective treatment option for auricular AVM.

4. Conclusions

We conclude that stereotactic radiosurgery is the effective treatment modality for auricular AVM. In our case, radiosurgery provided an excellent obliteration of the nidus and good cosmetic result eliminating the need for surgical reconstruction at the site of the nidus. The excellent outcome in our case warrants further study with larger sample size and longer follow up to validate the efficacy of stereotactic radiosurgery in treating auricular AVM.

Abbreviations

AVM	Arteriovenous Malformation
SRS	Stereotactic Radiosurgery
MRI	Magnetic Resonance Imaging
DSA	Digital Substraction Angiography
ECA	External Carotid Artery
GTV	Gross Tumor Volume
PTV	Planning Target Volume
V_{100}	Volume Receiving 100% of Prescription Dose
OAR's	Organs at Risk
QA	Quality Assurance
IMRT	Intensity Modulated Radiotherapy
CBCT	Cone Beam Computed Tomography
Dmax	Maximum Dose
Dmean	Mean Dose

Author Contributions

Mariya Deputy: Conceptualization, Data curation, Investigation, Methodology, Resources, Writing – original draft

Sanjay Hunugundmath: Conceptualization, Data curation, Investigation, Methodology, Resources, Supervision, Writing – review and editing

Amit Nirhali: Conceptualization, Data curation, Resources, Investigation, Methodology, Supervision, Writing – review and editing

Vishram Naik: Conceptualization, Data curation, Investigation, Methodology, Resources, Writing – review and editing

Sharad Gadhave: Conceptualization, Data curation, Investigation, Methodology, Resources, Writing – review and editing

Ethics Statement

Our case report didn't have any patient identifying information. Hence, didn't require ethics approval.

Funding

This work is not supported by any external funding.

Data Availability Statement

Not applicable.

Conflicts of Interest

The authors declare no conflicts of interest.

References

[1] Liu R, Chen J, Jia L, Pan B, Jiang H. Surgical management of auricular arteriovenous malformations: A literature review. Laryngoscope Investig. Otolaryngol. 2022 Apr; 7(2): 604-13. https://doi.org/10.1002/lio2.776

[2] Kim JB, Lee JW, Choi KY, et al. Clinical characteristics of arteriovenous malformations of the head and neck. Dermatol. Surg. 2017 Apr 1; 43(4): 526-33. https://doi.org/10.1097/DSS.0000000000000993

[3] Kohout MP, Hansen M, Pribaz JJ, Mulliken JB. Arteriovenous malformations of the head and neck: natural history and management. Plast. Reconst. Surg. 1998 Sep 1; 102(3): 643-54. https://doi.org/10.1097/00006534-199809030-00006

[4] Zou Y, Qiao C, Lin X, et al. Clinical course of extracranial arteriovenous malformations. J CraniofacSurg. 2020 Mar 1; 31(2): 372-6. https://doi.org/10.1097/SCS.0000000000006018

[5] Kansy K, Bodem J, Engel M, et al. Interdisciplinary treatment algorithm for facial high-flow arteriovenous malformations, and review of the literature. J. Craniomaxillofac. Surg. 2018 May 1; 46(5): 765-72. https://doi.org/10.1016/j.jcms.2018.03.002

[6] Fernández-Alvarez V, Suárez C, de Bree R, et al. Management of extracranial arteriovenous malformations of the head and neck. AurisNasus Larynx. 2020 Apr 1; 47(2): 181-90. https://doi.org/10.1016/j.anl.2019.11.008

[7] Vilela Chagas Ferreira M, Goldenberg DC, Kharmandayan V, Gemperli R. Management of arteriovenous malformation of the ear: a protocol for resection and reconstruction. Laryngoscope. 2020 May; 130(5): 1322-6. https://doi.org/10.1002/lary.28209

[8] Wu JK, Bisdorff A, Gelbert F, Enjolras O, Burrows PE, Mulliken JB. Auricular arteriovenous malformation: evaluation, management, and outcome. Plast. Reconst. Surg. 2005 Apr 1; 115(4): 985-95. https://doi.org/10.1097/01.PRS.0000154207.87313.DE

[9] Bulstrode NW, Lese I, Aldabbas M, Glover M, Robertson F, Rennie A. Arterio-venous malformations of the ear: description of distinct anatomical presentation and multidisciplinary management approach. J PlastReconstrAesthet Surg. 2021 Jul 1; 74(7): 1574-81. https://doi.org/10.1016/j.bjps.2020.11.018

[10] Wang D, Su L, Han Y, Fan X. Ethanol embolotherapy of high-flow auricular arteriovenous malformations with electrolytically detachable coil-assisted dominant outflow vein occlusion. Eur J VascEndovasc Surg. 2014 Nov 1; 48(5): 576-84. https://doi.org/10.1016/j.ejvs.2014.08.004

[11] Hua C, Jin Y, Yang X, et al. Midterm and long-term results of ethanol embolization of auricular arteriovenous malformations as first-line therapy. J VascSurg: Venous and Lymphatic Disorders. 2018 Sep 1; 6(5): 626-35. https://doi.org/10.1016/j.jvsv.2018.01.017

[12] Pham TH, Wong BJ, Allison G. A large arteriovenous malformation of the external ear in an adult: report of a case and approach to management. Laryngoscope. 2001; 111(8): 1390-4. https://doi.org/10.1097/00005537-200108000-00014

[13] Jin Y, Lin X, Chen H, et al. Auricular arteriovenous malformations: potential success of superselective ethanol embolotherapy. J. Vasc. Interv. Radiol. 2009 Jun 1; 20(6): 736-43. https://doi.org/10.1016/j.jvir.2009.02.008

[14] Zheng LZ, Fan XD, Zheng JW, Su LX. Ethanol embolization of auricular arteriovenous malformations: preliminary results of 17 cases. Am. J. Neuroradiol. 2009 Oct 1; 30(9): 1679-84. https://doi.org/10.3174/ajnr.a1687

[15] Yang F, Yang B, Qu Z, Tan Y, Lu F, Liao Z. Percutaneous ethanol sclerotherapy for auricular arteriovenous malformation: Our experience with 11 patients. Clin. Otolaryngol. 2020 Sep; 45(5): 811-7. https://doi.org/10.1111/coa.13567

[16] Whitty LA, Murray JD, Null WE, Elwood ET, Jones GE. An arteriovenous malformation of the external ear in the pediatric population: a case report and review of the literature. Can. J. Plast. Surg. 2009 Dec; 17(4): 45-7. https://pmc.ncbi.nlm.nih.gov/articles/PMC2827291/

[17] Kim SH, Han SH, Song Y, Park CS, Song JJ. Arteriovenous malformation of the external ear: a clinical assessment with a scoping review of the literature. Braz. J. Otorhinolaryngol. 2017 Nov; 83: 683-90. https://doi.org/10.1016/j.bjorl.2016.09.004

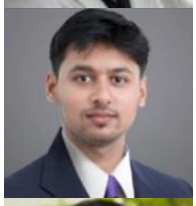
[18] Gupta A, Gupta S, Kumar A, Bhattacharaya S, Jha M, Tiwari V. High-flow vascular malformation of ear: a case report. World J. Plast. Surg. 2018 May; 7(2): 256. https://pmc.ncbi.nlm.nih.gov/articles/PMC6066713/

Biography

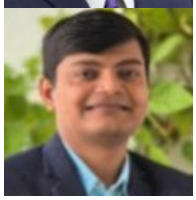
Mariya Deputy: Junior Consultant in department of Radiation Oncology at Sahyadrisuperspeciality hospitals, Pune, India. Special interest in Brain tumors, Breast cancer, Gynaecological cancers, SRS and SBRT.

Sanjay Hunugundmath: Lead Consultant in department of Radiation Oncology at Sahyadrisuperspeciality hospitals, Pune, India. He did his Masters in Radiation Oncology from Amrita Institute of medical sciences, Kochi. He completed his fellowship in stereotactic radiation drom National Cancer Centre, Singapore. He had extensive training in Clinical Oncology from TMC Kolkata.

Amit Nirhali: Chief Medical Physicist and RSO in department of Radiation Oncology at SahyadriSuperspeciality Hospitals, Pune, India. He has 15 years of extensive experience in commissioning Linac, Brachytherapy, Tomo-therapy machines, treatment planning, quality assurance of RT machines, setting up and designing radiotherapy departments. Key interest in intergrating machine learning, radiomics and AI in Radiation Oncology.



Vishram Naik: Senior Medical physicist in department of Radiation Oncology at Sahyadrisuperspeciality hospitals, Pune, India. 7 years of experience as a Medical Physicist with wide experience in treatment planning and quality assurance of RT. Key interest areas are Dosimetery and Advanced algorithm in Radiation Oncology.



Sharad Gadhave: Senior Radiation Therapist in department of Radiation Oncology at Sahyadrisuperspeciality hospitals, Pune, India. He did his advance diploma in Radiotherapy Technology from Tata Memorial Hospital, Mumbai. He ha wide experience in treating special procedure such as SRS, SBRT, DIBH, IGRT etc.

Research Field

Mariya Deputy: Oncology, Radiation Oncology, Molecular oncology.
Sanjay Hunugundmath: Oncology, Radiation Oncology, Molecular oncology.
Amit Nirhali: Medical Physics, Radiation Oncology, Artificial Intelligence in Radiation Oncology.
Vishram Naik: Medical Physics, Radiation Oncology.
Sharad Gadhave: Radiation Oncology.