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RADIOTHERAPY IN CUTANEOUS ANGIOSARCOMA OF THE SCALP: A CASE REPORT

Oncology/Radiotherapy

Dr. Diplu Choudhury — Assistant professor, Department of Radiation Oncology, State Cancer Institute, Gauhati Medical College, Guwahati, Assam.

Dr. Kaushik Baiswas* — Post Graduate Trainee, Department of Radiation Oncology, State Cancer Institute, Gauhati Medical College, Guwahati, Assam. *Corresponding Author

Dr Luri Borah — Assistant Professor, Department of Radiation Oncology, Nalbari Medical College and Hospital, Nalbari, Assam.

Dr Hima Bora — Registrar, Department of Radiation Oncology, State Cancer Institute, Gauhati Medical College, Guwahati, Assam.

Kalyanjit Dutta Baruah — Junior physicist, Department of Radiation Oncology, State Cancer Institute, Gauhati Medical College, Guwahati, Assam.

ABSTRACT

Background: Angiosarcoma of is a rare aggressive malignant tumour of vascular endothelial cells occurring more commonly in elderly males. Less than 5% soft tissue sarcomas occur in the head and neck, with only 10% termed as angiosarcomas. Presentation varies from small plaque to multicentric nodules. Surgery is the mainstay of treatment. Addition of adjuvant radiotherapy has definite benefit. Role of adjuvant chemotherapy is not yet well proven. **Case Report:** Here we are going to describe a case of Angiosarcoma of scalp involving the frontal and occipito-temporal region in a 62-year-old lady. The lady initially presented with a scalp swelling for 1 year for which she was evaluated at Surgical Oncology department, was diagnosed as a case of angiosarcoma of scalp, and eventually underwent wide local excision. Post OP HPE revealed margin positivity. After discussion in the tumour board, she was taken up for adjuvant radiation. She received a total dose of 66 Gy in 33 fractions through IMRT-IGRT technique using daily image guidance in helical tomotherapy machine. **Discussion:** After wide local excision, in margin positive tumours, the addition of adjuvant radiotherapy is beneficial in terms of improved local control and overall survival. For margin negative tumours, the benefit is still doubtful. Definitive radiation is recommended for unresectable or partially resectable tumours. Results with addition of adjuvant chemotherapy are better compared to radiotherapy alone. Historically whole scalp radiation technique has been used. The role of high dose brachytherapy with surface mould technique is there, Studies with use of newer techniques like helical tomotherapy are few. **Conclusion:** Angiosarcoma of the scalp is a very aggressive tumor with poor overall survival. Multimodal treatment including surgery and radiotherapy are effective in improving OS for patients with these tumours and addition of chemo or immunotherapy may further improve OS.

KEYWORDS

BACKGROUND

Angiosarcoma is an aggressive malignant tumour of the vascular endothelial cells occurring more commonly in elderly individuals.[1]

Overall, sarcomas are seen less frequently in the head and neck, constituting less than 1% of all head and neck malignancies.[2] In general, fewer than 5% soft tissue sarcomas occur in the head and neck, with only approximately 10% termed as angiosarcomas.[3]

The diagnosis of cutaneous angiosarcoma is challenging because it often presents insidiously as a bruise like lesion or purplish papule that may be mistaken for a benign lesion like haemangioma.[1] The tumour cells are located mainly in the dermis and may extend to the subcutaneous tissue. It spreads widely through the skin and recurs locally and metastasises early. Most common site of metastasis is lungs.[4]

Common risk factors associated with angiosarcoma are prior radiation exposure, chronic lymphedema (Stewart-Treves syndrome), and exposure to chemicals such as PVC, thorium dioxide, arsenic and radium.[1]

There are no definite criteria for staging of angiosarcoma. In the 8th edition of AJCC (The American Joint Committee on Cancer) staging manual for TNM (Tumor, Nodes, and Metastasis) staging, it has been excluded from soft tissue sarcoma chapter due to its infiltrative nature.[5]

On immunohistochemistry, vascular endothelial markers like CD (Cluster of differentiation) 31, CD 34, VWF (Von Willebrand factor)-related antigens are positive for this tumour. Also, lymphatic endothelial markers like D2-40, Prox-1, VEGFR (Vascular endothelial growth factor receptor) -3 are expressed signifying its lymphatic differentiation.[6]

Although overall prognosis of patients with angiosarcoma of the head and neck remains poor, with reported 5-year survival rate of approximately 10%[2]; surgery being the cornerstone of treatment; the use of adjuvant radiotherapy for better local control and OS (Overall survival) [7] and chemotherapy and immunotherapy with rIL-2 (recombinant interleukin-2)[4] have brought paradigm shift to the treatment of this uncommon tumour. Radiotherapy is also advocated for unresectable and incompletely resectable tumours.[5] Chemotherapeutic agents with effectiveness against angiosarcoma are anthracyclines and taxanes. But role of adjuvant chemotherapy is not yet proven.[1]

Here we are presenting a rare case of angiosarcoma of the scalp, treated for the first time in our institute, with surgical resection followed by adjuvant chemotherapy and adjuvant radiotherapy.

Case Report

A 62-year-old housewife from Jamunamukh, Hojai, Assam, presented with a scalp swelling for 1 year at outpatient department (OPD) of Surgical Oncology, State Cancer Institute (SCI), Gauhati Medical College, Guwahati on 08.03.22. The swelling was progressively increasing in size, especially in the last 1 week and there was occasional bleeding. Personal, past and family history were unremarkable with absence of any predisposing factor like prior radiation exposure, exposure to chemicals like PVC, Arsenic etc. On examination, her performance status (PS) was 1 ECOG (Eastern Cooperative Oncology Group) and general condition was fair. Local examination revealed (3x1.5) cm irregular swelling over frontal region of scalp. It was brownish in colour, firm in feel with irregular margins and there was bleeding upon palpation. There were also multiple small nodules, each of size of approximately 0.2 cm in occipito-temporal region. The rest of the scalp was normal. There were no palpable lymph nodes in the neck or anywhere in the body. CT (Computed Tomography) Brain done on 02.03.22 showed Ill-defined soft tissue thickening over the scalp in frontal region with normal underlying bones, suggestive of infiltrating vascular malformation of the scalp. MRI (Magnetic Resonance Imaging) Brain and Scalp done on 11.03.22 revealed a T2 mixed signal intensity, T1 and FLAIR (Fluid

Attenuated Inversion Recovery) isointense lesion in the scalp in the frontal region of size (11x5.4x1.1) cm. The lesion is irregular and shows restriction of diffusion in DWI (Diffusion weighted imaging) and ADC (Apparent diffusion coefficient) maps. There is post contrast enhancement.

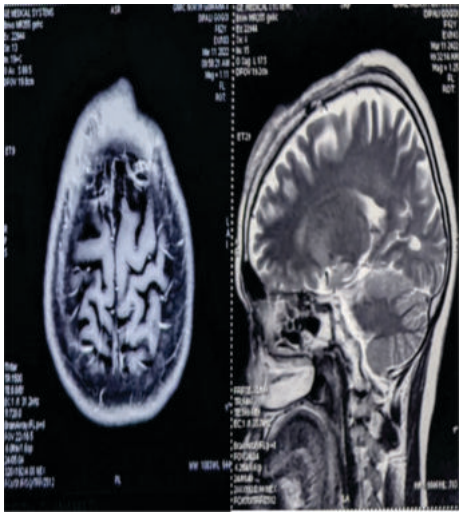

Fig 1 Preoperative T1 Weighted MRI Sequences In Axial & Sagittal Sections Showing The Lesion

HPE (Histopathological examination) of incisional biopsy of single greyish white soft tissue bits with attached skin and hair (scalp mass) reported on 14.03.22 revealed Poorly differentiated malignancy-Malignant melanoma/ Poorly differentiated squamous cell carcinoma. On IHC (Immunohistochemistry) (21.03.22), tumour cells express CD31, ERG and are negative for S100 and P40. KI 67 index is 35-40%. Impression- Angiosarcoma. CT Thorax and Abdomen done on 11.03.22 was within normal limit.

She then underwent Wide local excision (WLE) of the scalp mass and nodules under General anaesthesia with 1 cm margin on 07.04.22 at SCI. Defect coverage was done by split-thickness skin graft (STSG) from right thigh. Postoperative HPE (07.05.22) showed Poorly differentiated malignancy, WLE; site- scalp; size- 2x1.8x1 cm (largest nodule); LVI (Lymph vascular invasion)- seen; necrosis- not seen; margins- superior, inferior and medial margins free of tumour, lateral 0.2 cm away from tumour, deep margin involved by the tumour; pathological staging- pT_1N_x.

Consequently, she received adjuvant chemotherapy with Inj. Paclitaxel 3 weekly x 6 cycles and was referred to Radiation Oncology Department, SCI for adjuvant radiotherapy (RT).

At RTOPD, examination done on 14.10.22 revealed PS- 0 ECOG, wound healed, scar area healthy. There was no palpable lymphadenopathy. The case was then taken up for discussion in MDTB (Multidisciplinary tumour board) and there it was decided for adjuvant radiation.

Radiotherapy Technique

The patient was immobilised in supine position with a thermoplastic brain mould and CT simulation was done in SIEMENS® SOMATOM Definition AS-20 CT scanner. Images were acquired with a slice thickness of 3 mm. The images were then transferred to the treatment planning system in DICOM format and with proper image registration and fusion, contouring was done for CTV (Clinical target volume), PTV (Planning target volume) and OARs (Organs at risk) respectively. CTV included post operative tumour bed, preoperative gross tumour volume (GTV), along with a uniform margin of 4 cm circumferentially. The PTV was then created with a margin of 0.5 cm around CTV according to hospital protocol. The planning was done in PRECISION® software using IMRT-IGRT (Intensity modulated radiotherapy-image guided radiotherapy) technique with 14 non-co-planar beams to a dose of 66 Gy in 33 fractions in the RADIXACT® ACCURAY tomotherapy machine. Daily MVCT (megavoltage CT) was taken for setup verification as per our IGRT protocol. She successfully completed the prescribed radiation treatment. Treatment period was from 27.10.22 to 22.12.22.

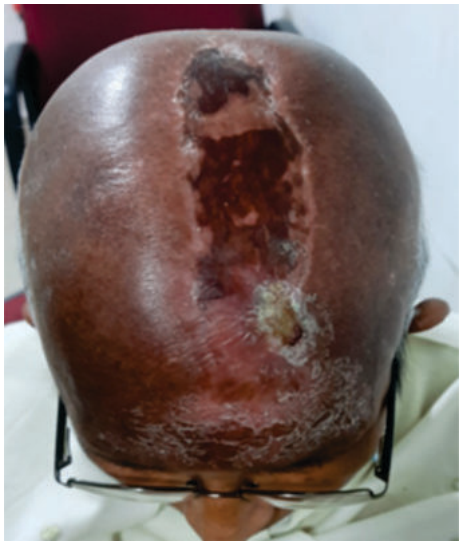

Fig 2 Post RT Scar

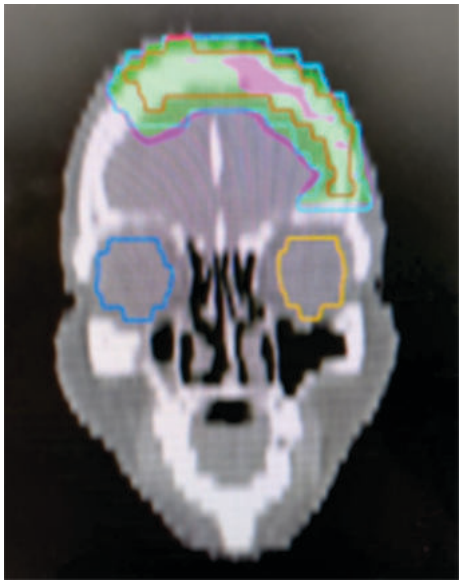

Fig 3 A: 100% (Green) And 98% (Pink) Isodose Coverage Of The Target Volume (Coronal)

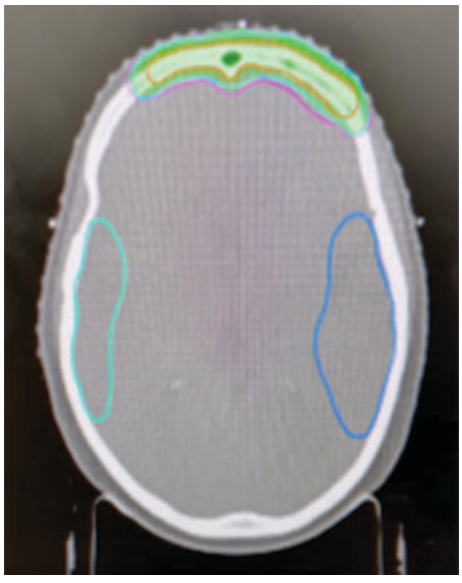

Fig 3 B: 100% (Green) And 98% (Pink) Isodose Coverage Of The Target Volume (Axial)

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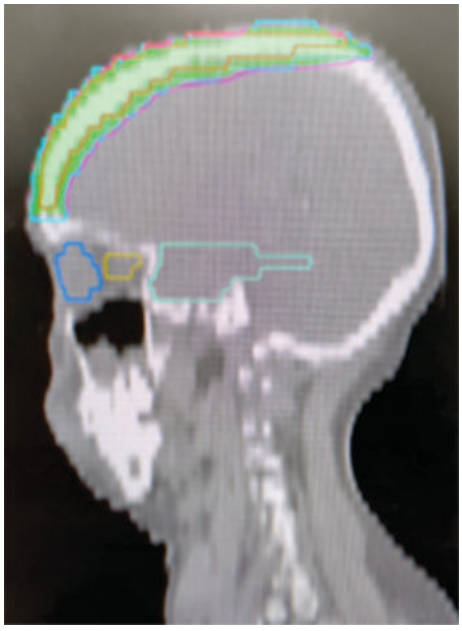

Fig 3 C: 100% (Green) And 98% (Pink) Isodose Coverage Of The Target Volume (Sagittal)

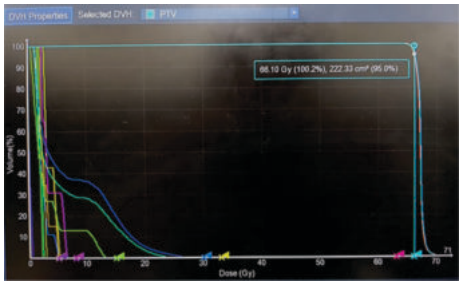

Fig 4 Dose Volume Histogram Showing The PTV Dose And Doses To The Oars (Check Fig 5 A And 5 B For Colour Codes Of Various Oars)

Dose Statistics Table | Dx Vx Values | Plan Information | Leaf Open Times | Dose Points

Name	Min (Gy)	Mean (Gy)	Max (Gy)	CI	nCI	HI	Coverage (%)
Patient	0.07	10.53	71.38	n/a	n/a	n/a	n/a
CTV	64.47	67.09	71.27	2.06	2.17	1.08	95.04
Rt eye	1.22	2.31	5.01	n/a	n/a	n/a	n/a
Lt eye	1.34	2.71	5.35	n/a	n/a	n/a	n/a
Rt ON	2.12	2.49	2.94	n/a	n/a	n/a	n/a
Lt ON	2.85	3.74	4.96	n/a	n/a	n/a	n/a
Rt Lens	1.67	1.94	2.25	n/a	n/a	n/a	n/a
lt lens	1.91	2.14	2.45	n/a	n/a	n/a	n/a
Chiasma	2.43	3.83	6.59	n/a	n/a	n/a	n/a
Brainstem	0.54	3.32	13.42	n/a	n/a	n/a	n/a
spinal cord	0.08	0.23	0.52	n/a	n/a	n/a	n/a
PTV	60.42	67.13	71.38	1.14	1.19	1.08	95.90
Rt Temporal lobe	1.08	6.25	26.41	n/a	n/a	n/a	n/a

Fig 5 A

Dose Statistics Table | Dx Vx Values | Plan Information | Leaf Op

Name	Dose (Gy)	Dose (%)	Volume (cm^3)	Volume (%)
PTV	62.70	**95.0**	234.04	100.0
PTV	66.00	**100.0**	224.55	95.9
PTV	69.30	**105.0**	1.79	0.8
PTV	70.62	**107.0**	0.26	0.1
PTV	69.21	104.9	**2.00**	0.9
Brainstem	13.42	20.3	0.00	**0.0**
spinal cord	0.52	0.8	0.00	**0.0**
Chiasma	6.59	10.0	0.00	**0.0**

Fig 5 B

Fig 5A And 5B: Dose Volume Statistics Table In Terms Of PTV And Oars

Follow up

After two months of treatment, she came for follow up with no specific complaints. Post OP scar was healthy and healed. There was no local recurrence or visible residual disease. The patient has been kept on follow up with consultation with surgical oncology team.

DISCUSSION

Angiosarcoma is a rare soft tissue sarcoma of vascular endothelial cells and may vary from well differentiated to those with significant anaplasia.[1,8] Though most soft tissue sarcomas are deep in location, angiosarcomas have a propensity for the skin and superficial soft tissue.[2] Most common predisposing factor is chronic lymphedema, but in a large number of patients there is no associated predisposing factor. In our patient also there was no identifiable risk factor.

Angiosarcoma occurs more commonly in elderly male of 68-76 years age group with male to female ratio being 2:1.[2, 3, 9] Our patient is a female, who is 62 years old.

Clinically, the lesion may be single or multifocal; bluish or violaceous; nodules, plaques, or flat infiltrating areas; and they can occasionally bleed or ulcerate. The lesions are usually painless (79.3%).[2]

Multifocality of the lesion is very much common in angiosarcoma and is associated with decreased disease-free survival. According to some authors, this is due to delayed diagnosis of angiosarcoma, which allows the lesion to proliferate unopposed, resulting in eventual poor prognosis of the tumour. Median delay of diagnosis is 5.1 months.[2, 10, 11] Another poor prognostic factor is advanced age.

Three histologic patterns are seen microscopically in angiosarcoma, namely vascular channels, sheets of cells, and cells with undifferentiated morphologic features.[2] But histologic grade does not affect prognosis.[12, 13] Stage is rather a more important predictor of survival, with patients having tumours <5 cm in greatest dimension have a significantly better prognosis compared with patients who have larger lesions.[14]

Due its multicentric nature and extensive microscopic spread, wide excision of angiosarcoma is done with an aim to achieve histologically negative margin followed by staged reconstruction with split thickness graft, local flaps or free flap.[8] In our patient also wide local excision with 1 cm margin was done followed by STSG repair from right thigh.
An important prognostic factor for angiosarcoma of scalp is whether the patient has received radiotherapy or not.[2] Patients receiving radiation therapy as adjuvant treatment has a median survival of almost 4 times longer than patients receiving no radiation therapy.[2] It is offered historically as postoperative low-dose, very wide-field radiation after excision of clinically evident tumour.[15] Diffuse multifocality is another indication of radiotherapy. Advantage of radiotherapy, however, is not well proven in case of margin negative tumours. Our case had margin positivity in her post operative histopathology. In case of extensive tumour, high dose brachytherapy with surface mould technique can also be used. Studies showing results with treatment using tomotherapy are rare. In the adjuvant setting, for negative margins, dosage ranges from 45 to 60 Gy in 25 to 30 fractions. For positive margins dosage ranges from 60 Gy to 66 Gy in 30 to 33 fractions.[1]

Definite radiotherapy is employed for unresectable or incompletely resectable tumours. Treatment is delivered using a two-step cone-down technique with phase 1 CTV including the entire scalp receiving a median dose of 50 Gy in 25 fractions followed by a dose of 20 Gy in 10 fractions to initial tumour site with additional margins. But results with radiotherapy with adjuvant chemotherapy are better compared to radiotherapy alone.[1,5]

One of the novel modalities of treatment of angiosarcoma of scalp is use of intralesional interferon alpha-2b and interferon-2 with surface radiotherapy in lieu of surgery.[16] The role of conventional chemotherapy, however, is still debatable because of the rarity of this tumour. Paclitaxel has been widely used recently, due to its antiangiogenic properties.[17]

CONCLUSION

Angiosarcoma of the scalp is an aggressive rare tumour with poor prognosis. Presence of good prognostic factors like younger age, fewer

lesions at diagnosis, small tumour size, ability to obtain clear margins along with definitive treatment with surgery and radiation provides best chance of cure. Newer techniques of radiation like tomotherapy are to be explored more in future with randomized trials for better local control and overall survival of this rare tumour with sparing of critical organs and improved quality of life. The role of chemotherapy is also to be established with optimal chemotherapy regimen for systemic control. Lifelong surveillance is recommended to detect any delayed distant metastasis.

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