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External beam radiotherapy of inoperable wilson-jones angiosarcoma in scalp as monotherapy: partial scalp irradiation with volumetric modulated arc therapy

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ABSTRACT

Angiosarcomas of the scalp are a rare and highly aggressive malignant tumor with a poor prognosis. External Beam Radiation Therapy (EBRT) alone has been rarely used for upfront treatment. We hereby present a case of an elderly gentleman with an inoperable scalp lesion (Wilson-Jones AS) with multiple comorbidities precluding surgery and chemotherapy, and was therapeutically approached with external beam radiation therapy using Volumetric Modulated Arc Therapy (VMAT). A brief course of management and clinical context are discussed herein.

Key words: beam radiotherapy, monotherapy, Volumetric Modulated Arc Therapy (VMAT).

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INTRODUCTION

Angiosarcoma (AS) are relatively rare locally aggressive lesions with poorly understood etiological factors and unknown clinical prognostication. As such, an individual clinical presentation requires a multidisciplinary approach, as no definitive guidelines exist. External Beam Radiation Therapy (EBRT) alone has been rarely used for upfront treatment. We hereby present a case of an elderly gentleman with an inoperable scalp lesion (Wilson-Jones AS) with multiple comorbidities precluding surgery and chemotherapy, and was therapeutically approached with External Beam Radiation Therapy (EBRT) using Volumetric Modulated Arc Therapy (VMAT).

CASE PRESENTATION

84 years old with Coronary Artery Disease, post Percutaneous Transluminal Coronary Angioplasty (PTCA), Diabetes Mellitus 2 (DM2), presented with a right sided scalp lesion of six months duration. Clinically, it extended to the nape of the neck towards the right occipital region and external pinna with involvement of the temporal region. On histopathology. It was diagnosed as Angiosarcoma. 18-Flouro-DeoxyGlucose (FDG) Positron Emission Tomography (PET-CT) showed an ill-defined soft tissue thickening over right parietal and temporal regions, extending from the vertex to the right preauricular region over the posterolateral aspect of the right posterior parietal region, measuring 0.8 cm × 4 cm × 3.5 cm (SUV_{max} 12.9). After multidisciplinary input with consideration of co-morbidities and high risk of surgery, the patient's refusal for definitive surgery was documented.

Dosimetric parameters and treatment

Patient was immobilised using 3 clamps, and simulation was performed with a carbon fibre table top 16 slice positron emission tomography CT-simulator (Simens® Biograph Truepoint® HD, Siemens AG, Medical Solution, Erlagen, Germany). External fiducial markers were placed all along with the clinical extent in pre-treatment simulation CT scan with 1.0 mm inter-slice thickness. They were generated with DICOM format and transferred to treatment planning system (TPS). Tumour volumes such as GTV (defined on FDG-PET and CT based thickening), CTV (with a margin on 1.5 cm-2 cm), isometrically expanded to avoid critical OAR's and expanded from the limits of fiducials marking the clinical extent on palpation) and Planning Target Volume (PTV) (3 mm as per the institutional protocol)

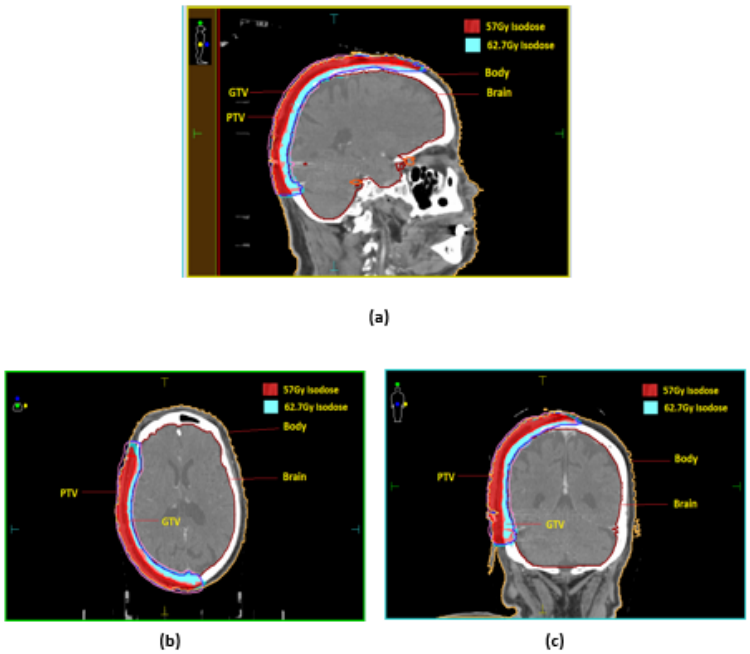

Fig. 1. (a, b, c). Isodose distribution of Angiosarcoma in scalp using volumetric modulate arc therapy

and critical Organ At Risk volumes (OAR) were delineated.

The Volumetric modulated arc therapy (VMAT) plan was generated for Elekta Synergy™ linear accelerator (Elekta Ltd, Crawley, UK) with 1 cm leaf width at isocenter. In addition, planning parameters of 3.0 mm grid size, 2% statistical uncertainty and 6.0 mm segment width were used in Monaco™ V.5.1 TPS. Two different partial arcs, such as ARC-1 180°/75° and ARC-2 180°/195° 20° gantry interval, were used for plan generation. VMAT plan was analysed using Dose Volume Histogram (DVH) generated by the Monaco™ TPS.

EBRT (using VMAT) was delivered to a total dose of 66Gy/30 fractions to Gross Tumour Volume (GTV) at 2.2Gy/fraction, and 60Gy/30 fractions to Clinical Target Volume (CTV) at 2Gy/fraction to PTV by VMAT to 95% isodose coverage as shown in figure 1. Patient developed Grade III dermatitis during the course of radiation therapy, which was approached conservatively with topical and oral antibiotics/antifungals, topical mild steroid ointments and WHO Step I analgesia. He had a gap of 14 days, and 1 fraction was added to the overall dose of administered treatment schedule.

DISCUSSION

AS (cutaneous variant accounts for less than 1% of all sarcomas) can arise from any soft tissue with an aggressive clinical course characterised by high rates of local recurrence. They represent 15% of all head and neck sarcomas, with the most common clinical presentation on the scalp and gender predisposition towards men.

It has multifactorial aetiology with a combination of environmental, occupational, familial or iatrogenic factors. Prior radiation exposure or chronic lymphoedema, especially in post mastectomy radiation therapy (Stewart-Treves syndrome), putative exposure to vinyl chloride, arsenic and thorium dioxide (Thorotrast) has been previously implicated. It may have a syndromal association with Neurofibromatosis and Maffucci, and rarely present as heterologous components of benign or malignant nerve sheath tumours. When it presents on the face or scalp of elderly patients, it is called Idiopathic or Wilson-Jones angiosarcoma. Its 5-year Overall Survival (OS) rate is only 10%-15% because of its tendency for early haematogenous metastasis to the lungs.

Histopathologically, AS presents with variable morphologic appearance, ranging from cytologically bland to solid sheets of poorly demarcated highly pleomorphic cells with irregular anastomosing vascular channels lined by atypical endothelial cells ramifying through dermal collagen and subcutaneous soft tissues. The latter variant may present in poorly differentiated angiosarcoma, often with intratumoral haemorrhage. Tumor cells are pleomorphic with either spindle, polygonal and epithelioid shapes forming papillae or solid nests within vascular lumina and mitotically active. Histologic grade is irrelevant in prognostication, with no definitive biomarkers. Predominant solid pattern in head and neck angiosarcomas may instead portend a favourable prognosis. Presence of necrosis, epithelioid morphology, and depth of invasion are other factors determining clinical outcomes [1-4].

On Immunohistochemistry (IHC), AS positively expresses endothelial cell markers (CD31, CD34, ERG, FLI1, VEGF and factor VIII), variable expression of lymphatic marker podoplanin and negative for HHV8 (Human Herpesvirus 8). Epitherloid variant may express cytokeratin, EMA and CD30. Genotypically, they have an up-regulation of vascular specific receptor tyrosine kinases (TIE1, KDR, TEK and FLT), recurrent PTPRB and PLCG1 mutations, and overexpression of HIF1 alpha, HIF2 beta (upstream regulators of VEGF) involved in angiogenesis. c-MYC gene amplification is observed in radiation induced AS. FLT4 amplification has been detected in 25% of secondary angiosarcomas.

Clinically, it is locally aggressive, presenting as a rapidly growing macular, nodular or plaque (classically multicentric, indistinct

borders showing hemorrhagic ulceration; easily bruised, purpuric area) with a high propensity for nodal and systemic metastases. Differentials include capillary haemangiomas, Kaposi sarcoma, epithelioid hemangioendotheliomas, and hemangiopericytomas. 10% of patients in the head and neck region may have clinically overt cervical lymph-nodes. Magnetic Resonance Image (MRI) reveals local extent with post-contrast enhancement as characteristic. There is an emerging role of 18Flouro-DeoxyGlucose (18FDG) Positron Emission Tomography (PET) both in local and systemic assessment. The most common site of metastasis is the lung, followed by the lymph nodes, bone, and liver.

AS requires multimodal approaches without a clear consensus due to its rarity. Surgical excision with clear wide margins (R0 resection) remains the mainstay, followed by local radiotherapy alone. R0 resection is difficult in view of multifocality, and extensive spread often complicates reconstructions. Neoadjuvant chemotherapy may be used preoperatively if the disease involves the nose or the periorbital region, with the aim of bulk reduction and simplifying surgical resection [2]. Doxorubicin based regimens with Progression-Free Survival (PFS) of 3 months are recommended in patients with systemic metastases. Paclitaxel shows a better efficacy compared with doxorubicin, with a median PFS of 4-5 months. Anti-angiogenic drugs (sunitinib, sorafenib, thalidomide) are no longer used. Immunotherapy is emerging as a newer treatment option for patients with local progression or in recurrent/relapsed scenarios. Targetted Therapy (anti-angiogenic agents against VEGF) using bevacizumab has been proposed as monotherapy or in combination with EBRT or systemic chemotherapy. Other antiangiogenic agents have also been studied (sorafenib, sunitinib or pazopanib). When treated as part of a multidisciplinary approach, the 5-year survival rate may reach up to 62% [5].

Adjuvant EBRT to a dose of 60 Gy in 30 fractions for primary tumor improves local control and OS. In case of inoperable patients, a higher dose of 70 Gy in 30 fractions has been reported (through mild hypofractionation or Simultaneous Integrated Boost- SIB) [6, 7].

CONCLUSION

Angiosarcomas of the scalp are a rare and highly aggressive malignant tumor with a poor prognosis. Because of the rarity of this disease, the optimal management strategy has not yet been established. Wide local excision combined with postoperative radiotherapy is an optimal treatment, although our experience shows that EBRT as monotherapy for inoperable elderly patients is a feasible alternative. Chemotherapy is indicated in palliation alone for systemic metastases.

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