

Radiation-Induced Sarcoma Post Radiotherapy Treatment of Nasopharyngeal Carcinoma

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ABSTRACT

Introduction: Nasopharyngeal carcinoma (NPC) is a squamous cell carcinoma that commonly arises from the Fossa of Rosenmuller. The primary treatment for NPC is radiotherapy, which in long term is known to induce secondary malignancies. Radiation-induced sarcoma (RIS), a late complication of radiotherapy, is a rare tumour with a reported incidence of 0.03-0.8%. Prognosis is poor with a 5-year survival rate of 30%.

Report: Our case is a 65-year-old lady who has completed concurrent chemotherapy and radiotherapy 15 years ago. She presented with 1 month history of painful swelling over the right temporomandibular joint. Initially she was treated as parotitis with a differential of osteoradionecrosis based on orthopantomograph findings. Her swelling increased in size (6x8 cm) and started complaining of trismus and persistent headache. Biopsy was reported as high grade epithelioid sarcoma. CECT neck showed a large heterogeneously enhancing right masticator space mass with intracranial extension. In view of the extent of the disease, patient was counselled for palliative chemotherapy.

Conclusions: The rarity of RIS poses a diagnostic and treatment challenge to clinicians. Early clinical investigation can be difficult due to induration and fibrosis of the irradiated fields. Although new techniques of radiotherapy have been used to minimize the radiation field, outcome of studies comparing new techniques against conventional radiotherapy on incidence of secondary malignancies remains debatable. Studies have shown neither additional radiation nor chemotherapy will improve overall survival. With the improved treatment regime for NPC and variability of RIS latency period, this necessitates lifelong monitoring of NPC patients post radiotherapy.

Keywords: Nasopharyngeal carcinoma, radiation-induced sarcoma, radiotherapy

INTRODUCTION

Nasopharyngeal carcinoma (NPC) is a squamous cell carcinoma that commonly arises from the fossa of Rosenmuller. The primary treatment for NPC is radiotherapy. Ionizing radiation, an important modality in the treatment of head and neck cancer has been known to induce secondary malignancy in the long term, and squamous cell carcinoma is the most reported pathology. ^[1] Radiation-induced sarcoma (RIS), arising post treatment of NPC, is a rare tumour entity with a reported incidence of 0.03-0.8%. ^[2] The latency period for RIS

is between 6 to 25 years. With the current advancement in treatment of NPC, the incidences of RIS may increase due to improved survival rate.

BACKGROUND

Our case is a 60-year-old lady with a history of NPC. She was diagnosed 15 years ago and completed her concurrent chemotherapy and radiotherapy without complications. However, she started to complaint of a painful swelling over the right temporomandibular joint region for 1 month. Initially it was treated as parotitis

with a differential of osteoradionecrosis. Her condition worsens with the swelling gradually extends to involve the right temporal region. Patient also complaints of trismus, otalgia and persistent headache. On clinical examination, the mass measures 6 x 8 cm and extends from the right temporal region inferiorly to the angle of mandible. Patient has right sided facial numbness but denies any weakness. Enlarged lymph node was palpable at right level Ib. Initial fine needle biopsy failed to yield results. An incision biopsy was done and reported to be an intermediate to high grade epithelioid sarcoma. CECT neck was reported as an aggressive large heterogeneously enhancing right masticator space mass with base of skull and intracranial extension (Fig 1). Due to the extension and aggressive nature of the disease, patient was counselled for palliative chemotherapy. She was started on doxorubicin but unfortunately patient succumbed to her disease a few weeks later.



Fig 1: CECT Neck showing a large heterogeneously enhancing right masticator space mass with bony erosion and extension to the base of skull

DISCUSSION

The rarity of RIS post treatment of NPC presents a diagnostic and treatment challenge to clinicians. Radiation induced tumours usually involve the skin, thyroid gland and salivary glands. The presentation of RIS may mimic that of radiation osteitis and cases of misdiagnosis have been reported. [2] In our case, the initial orthopantomograph was consistent with osteoradionecrosis (Fig 2).



Fig 2: Ortopantomograph showing radiolucency at right mandible extending from the coronoid process to the sigmoid notch

The early clinical presentation of RIS may be difficult due to induration and fibrosis of the irradiated fields. This may explain the failure to obtain a positive result during the initial fine needle biopsy. The latency period between RT and RIS ranges between 6 to 25 years. Although RIS can be attributed to complications of irradiation, it is almost impossible to differentiate it from a primary sarcoma as the lower limit of the latency period is still controversial. [2] Patients with NPC normally receive a radiation dose of about 62.5 Gy. This put them at higher risk as it has been shown that a total dose of 55Gy or more increases the incidences of RIS. [1] Although new techniques of RT have been used to reduce the radiation dose to surrounding organs, the outcome of the recent studies comparing the new techniques against conventional RT on the incidence of secondary tumours remained debatable. [3]

Prognosis for RIS of the head and neck is poor with a 5 year disease free survival rate of 8%. It is hampered by the limitations for further radiotherapy and the relative insensitivity of the tumour to chemotherapy. [4] Currently, a complete surgical resection of the tumour is the mainstay of treatment. However, due to the anatomical relation of the tumour to the base of skull and other vital structures, resection of the tumour with a proper margin is technically very difficult. A multivariate analysis by Xi et al in their

study showed that a complete surgical resection is the only independent predictive factor for better survival. [2]

CONCLUSION

RIS is a rare oncologic tumour and has a poor prognosis. With its aggressive nature and poor sensitivity to chemotherapy, a complete surgical resection seems to be the treatment of choice [5] Current improved treatment regime for NPC will see an increase in the incidence of RIS. Therefore, suspicions level for RIS should be high in patients who present with a secondary mass post treatment. The widely variable latency periods of RIS necessitate lifelong monitoring of NPC patients who have undergone RT, especially in young patients.

Conflict of Interest

Sin Wee Lim and Tengku Mohamed Izam Tengku Kamalden declare that they have no conflict of interest

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