Head and Neck Radiation-Induced Sarcoma: Report of Two Cases

Case Report

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ABSTRACT

The incidence of radiation-induced sarcoma (RIS) in head and neck region is increasing in trend nowadays. There are few cases have been reported. However, the exact aetiology of radiation-induced sarcoma (RIS) is still unknown. We report two cases of radiation-induced sarcoma (RIS) which the primary malignancy were basal cell carcinoma of the cheek and nasopharyngeal carcinoma. These patients had received a total of 70Gy and 65Gy radiotherapy for 13 years and 6 years back respectively. Histopathological examinations for both of these patients revealed a spindle cell sarcoma and a high-grade sarcoma respectively. Both cases were inoperable in view of advanced disease at presentation. We will discuss on the clinical findings, histopathological findings and the main characteristic of radiation-induced sarcoma (RIS) in imaging. The dilemma of managing radiation induce sarcoma in head and neck patient will also be discussed as managing previously radiated head and neck primary is undeniable challenging

Key Words: Head and neck RIS, radiation-induced sarcoma, sarcoma post radiation.

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INTRODUCTION

The earliest description of radiation-induced sarcoma (RIS) was reported in 1936 by Warren and Sommer[1] Until today, the number of reported cases is increasing. Over the years, radiation therapy has been driven by constant technological advances and approximately 50% of all patients with localized malignant tumours are treated with radiation at some point in the course of their disease. Thus it would be expected to have an increase probability of radiation induce tumour. RIS following radiotherapy for NPC is less than 0.5% while other head and neck cancer has yet to be reported[2] Nevertheless, the overall incidence of RIS in a lifetime is about 0.06-0.3%[3]

CASE REPORT

Case 1:

A 24-year-old lady was diagnosed as nasopharyngeal carcinoma 13 years ago, and received a total dose of 70 Gy, 6MV photon energy of external beam radiation therapy with four cycles of cisplatin therapy. currently presented with progressive painless right neck swelling for four months duration. It was associated with loss of appetite and loss of weight. She has no difficulty in breathing or dysphagia. No intracranial or cranial nerve involvement. Other systemic reviews were unremarkable.

Examination revealed a non-tender swelling located at right level II extending to level IV measuring approximately 7.0 x 8.0 cm (Figure 1). The mass was firm-to-hard and fixed to the underlying structure. The cranial nerves were all intact bilaterally.

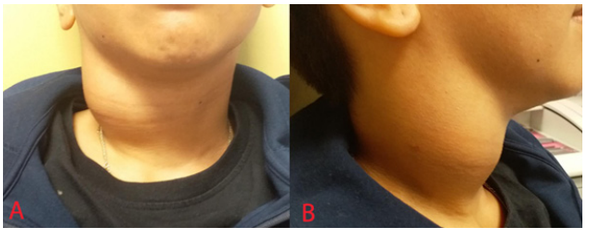

Fig. 1: The non-tender, hard right level II swelling, extending down to level IV measuring about 7.0 x 8.0 cm

Magnetic resonance imaging (MRI) of the neck showed right neck mass measuring 5.6 x 4.8 x 8.9 cm with engulfment of the right common carotid artery and more than 180° encasement at proximal part of right internal carotid artery. An ultrasound-guided trucut biopsy confirmed a leiomyosarcoma.

She was staged as T4bN0M0 and surgical resection was not proceeded as the tumour have involved the right common carotid artery and right paraspinal muscle. Thus,

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she was offered for chemotherapy. She had undergone 1 cycle of chemotherapy but unfortunately, she succumbed to the disease.

Case 2:

A 65-year-old man who was previously radiated for right nasal nodular basal cell carcinoma 6 years age. He had received a total dose of 65 Gy , 6MV photon energy of Intensity Modulated Radiation Therapy technique. It was radiated at faciocervical site using linear accelerator machine. He currently presented with right nasal and cheek swelling and diplopia for 7-month of duration. It was associated with dysphagia, loss of appetite and weight. He was a mouth breather as the mass has occluded his nostril.

Examination revealed a foul-smelling fungating mass over the right nostril involving the septum and ulceration on the right side of the hard palate. There was olfactory, oculomotor and trigeminal nerve palsy. Other cranial nerves were intact bilaterally. The systemic reviews were unremarkable.

Contrast-enhanced computed tomographic (CT) scan of the neck and paranasal sinuses showed heterogeneously enhancing mass with central hypodensity occupying the entire right maxillary sinus, extending anteriorly to subcutaneous tissue of right cheek (Figure 2). The mass extended enormously to few areas which are the right nasal cavity, bilateral frontal and sphenoid sinus, right orbit (muscles), oral cavity and right medial pterygoid muscle. Overall the mass had invaded almost all the bony structure in right oral, nasal cavity and paranasal sinuses.

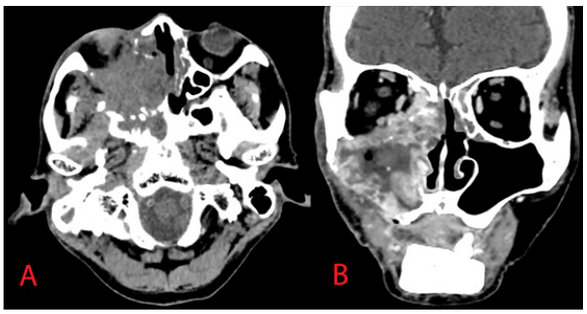

Fig. 2: Contrast-enhanced CT paranasal sinuses of axial (A) and coronal (B) section showed heterogeneous enhancing mass had invaded almost all the bony structure in right oral, nasal cavity, paranasal sinuses as well as the right orbit.

Histopathological examination showed malignant cells with positive immunohistochemical stain for h-Caldesman which was consistent with high-grade sarcoma. The tumour was staged as T4aN1M0 inoperable thus he was referred for radiotherapy and chemotherapy. Unfortunately, he refused any treatment and succumbed to the illness after five months.

DISCUSSION

The incidence of radiation-induced sarcoma (RIS) ranges from 0.03 to 0.3% and the most common location was the maxillary region, including the maxillary sinus, alveolar process, palate, and adjacent nasal cavity[2,4] RIS developed within the radiation volume for the primary tumour in which the mean radiation dose for the primary tumour was 33 Gy (range: 25–50Gy); the mean latency period between radiation therapy and occurrence of the sarcoma was 15 years (range of 3–50 years)[5]

For bone sarcomas Cahan et al. has developed the first set criteria for RIS[6] Murray et al. revised these to include other soft tissue sarcomas[7] These criteria was further modified as such: (i) a prior history of radiation; (ii) development of a sarcoma in the field of radiation; (iii) a latency period of at least 2 years between radiation and appearance of the sarcoma and (iv) evidence that the sarcoma is histologically different from the irradiated primary cancer[8]

The average latency period from the time-of-radiation to the time-of-diagnosis of the RIS was 12.6 years (range of 3–30 years) and it was believed that squamous cell carcinoma will be presented at a mean duration of 12.5 years post radiation. Sarcomas will be presented a bit sooner, around 8.8 years post radiation.[5] It was suggested that patient with head and neck cancer should need long term follow up at least for 15 years. Both our patients have been exposed to the mean radiation which has been mention above and they presented after 13 years and 6 years respectively. Thus, long term surveillance follows up is necessary for post radiation patients.

The common symptoms depend on related area of RIS. Those with RIS at maxillofacial region usually presented with painless or painful swelling associated with local or intracranial symptoms[5] As for our first patient, she presented with painless nasal mass with cranial nerve palsy, whereas for the second case he presented with painless neck swelling with no other symptoms.

The main characteristic of RIS on CT and MRI was soft tissue mass appearance. All cases showed heterogenous density before and after intravenous administration of contrast. however, there is no pathognomonic findings for MRI as the imaging appearances is variable, but the tumour will demonstrate aggressive features, and some appear benign which can lead to misdiagnosis[9]

The management for RIS is complete surgical resection of the tumour and it is now generally believed that it provides the only chance of a cure[5,10] Reports revealed that cases of RIS are considered radioresistant[11,12] However, study by Walter on the effect of sensitivity of tumour based on radiation and sensitizing agent shows

that the radio responsiveness of sarcomas is very variable and dependent on histology, kind of radiation, and various concomitantly given drugs[13] He also conclude that the rate of complete sustained remissions by radiation therapy alone or in combination with drugs is still far from satisfactory although progress has been made through the use of sensitizing agents thus complete surgical resection could be helpful. This is why managing RIS is very challenging.

The prognosis of RIS often site-related, reflecting the fact that surgical resection offers the only hope of cure. Unfortunately, they often occur in unfavourable sites, diagnosed late and poor response to therapy[14] The above cases were inoperable and both patients did not survive due to advance disease.

CONCLUSION

Radiation-induced sarcoma (RIS) is an aggressive tumour with poor survival rate as most of the patients presented late. Radical surgical resection with clear margin is still the gold standard of treatment. Although RIS is not preventable, but early detection can increase the survival rate. The management of previously radiated head and neck primary is undeniable challenging.

CONFLICT OF INTEREST

The authors declare no conflict of interest

REFERENCE

1. Warren S, Sommer G.N. Fibrosarcoma of the soft parts with special reference to recurrence and metastasis. Arch Surg. 1936;3: 425–50.

2. Abrigo Jill, King Ann, S.Leung. MRI of radiation-induced tumors of the head and neck in post-radiation nasopharyngeal carcinoma. Eur Radiol. 2009;19:1197–205.

3. Rosko A.J, Birkeland A.C, Chinn S.B, Shuman A.G, Prince M.E, Patel R.M et al. Survival and margin status in head and neck radiation-induced sarcomas and de novo sarcomas. Otolaryngol Head Neck Surg. 2017;157: 252–9.

4. Cahan WG, Woodard HQ. Sarcoma arising in irradiated bone; report of 11 cases. Cancer. 1948;1:3–29.

5. Murray EM, Werner D, Greeff EA, Taylor DA. Postradiation sarcomas: 20 cases and a literature review. Int J Radiat Oncol Biol Phys1999; 45:951–61.

6. Laskin WB, Silverman TA, Enzinger FM. Postradiation soft tissue sarcomas. An analysis of 53 cases. Cancer. 1988; 62:2330–40.

7. Korampalli TS, Mathew B, Staffor ND. Post radiation myofibrosarcoma of hypopharynx. J Surg Case Reports. 2013;2013:49–51.

8. Yang Q , Mo Y, Zhao Q, Ban X, He M, Cai P et al. Radiation-induced sarcomas of the head and neck in post-radiation nasopharyngeal carcinoma. Radiol Medica. 2017;122:53–60.

9. Debnam JM, Guha-Thakurta N, Mahfouz YM, Garden AS,Benjamin RS, Sturgis EMet al. Radiation-associated head and neck sarcomas: Spectrum of imaging findings. Oral Oncol. 2012; 48:155–161.

10. King AD, Ahuja AT, Teo P, Tse GMK, KewJ. Radiation induced sarcomas of the head and neck following radiotherapy for nasopharyngeal carcinoma. Clin Radiol.2000; 55:684–9.

11. Mavrogenis AF, Pala E, Guerra G, Ruggieri P. Post-radiation sarcomas. Clinical outcome of 52 Patients. J Surg Oncol. 2012;105:570–6.

12. Lagrange JL, Ramaioli A, CHateau MC, Marchal C, Resbeut M, Lagarde P et al.Sarcoma after radiation therapy: Retrospective multiinstitutional study of 80 histologically confirmed cases. Radiology.2000;216:197–205.

13. Rhomberg W. The radiation response of sarcomas by histologic subtypes: a review with special emphasis given to results achieved with razoxane. Sarcoma.2006;2006: 1-9.

14. Chan, JYW, To VSH, Wong STS, Wei WI. Quality of dying in head and neck cancer patients: The role of surgical palliation. Eur Arch Oto-Rhino-Laryngology. 2013; 270:681–8.