Radiation-induced high grade spindle cell sarcoma of the sternomastoid muscle: case report

Zaid Hamdoon¹,2,3
Waseem Jerjes¹,4,5
Raed Al-Delayme¹
Tahwinder Upile⁴
Fancis Vaz⁶

(1) Department of Oral and Maxillofacial Surgery, School of Dentistry, AL-Yarmouk University College, Baghdad, Iraq
(2) Unit of Oral and Maxillofacial Surgery, UCL Eastman Dental Institute, London, UK
(3) Department of Oral and Maxillofacial Surgery, University of Mosul, Mosul, Iraq
(4) Department of Surgery, UCL Medical School, London, UK
(5) Leeds Institute of Molecular Medicine, University of Leeds, Leeds, UK
(6) Head and Neck Centre, University College London Hospitals, London, UK

Corresponding author

Zaid Hamdoon

and

Francis Vaz

ZH: zaid19772000@yahoo.com
WJ: waseem_wk1@yahoo.co.uk
RA: raedmaxfax@yahoo.com
TU: mrtupile@yahoo.com
FV: francis.vaz@uclh.nhs.uk
Abstract

Sarcomas developing as primary malignancies of the head and neck are a rare complication after radiation therapy. This kind of sarcoma has variable clinicopathological appearances and behaviour. Radiation-induced spindle cell rhabdomyosarcoma (RMS) of the sternomastoid muscle is a very rare sarcoma and has very seldom been described in the literature.

Herein we report the development of a rapidly growing mass over the lateral side of the neck appearing after 7 years in a patient with a history of laryngeal carcinoma who received radiotherapy. The process of diagnosis and management using combined surgery and targeted brachytherapy are discussed.

The patient experienced discomfort and oozing of the wound up to 2 months after surgery; however, complete response with satisfactory adaptation and shrinkage of the pectoralis major pedicled muscle flap occurred after 1 year. The patient is currently disease free 5 years postoperatively.

Radiation-induced spindle cell rhabdomyosarcoma of the sternomastoid muscle after treatment for laryngeal carcinoma may occur a long time after radiation therapy and be clinically aggressive, radiographically distinctive, and require multidisciplinary management.
Introduction

Although radiotherapy is known to be an effective treatment modality in the management of malignant disease, ionizing radiation can also induce malignant tumour formation (1-8). This has been shown by studies on the sequelae of human exposure to radiation from atomic testing. The first recorded case of a radiation-induced cancer was a squamous cell carcinoma that arose in the hand of a 33-year-old technician who had been testing roentgen tubes for 4 years (2).

Radiation-induced sarcoma (RIS) of the head and neck is a rare and long-term complication of treatment with radiotherapy. Although there are many case reports on RIS in the medical literature, these cases cannot provide reliable information on outcome of each pathology as there are few reported series published with only a limited number of cases.

Post-irradiation sarcoma (PIS) is another name given to RIS, with histological types including osteosarcoma, fibrosarcoma, angiosarcoma and malignant schwannoma. Spindle cell rhabdomyosarcoma (RMS) is a rare PIS first described in children in 1992 as a neoplasm composed mainly of fascicular spindle-shaped cells that show immuno-histochemical and ultrastructural evidence of myogenic differentiation (1). The aetiology of spindle-cell RMS in adults is unknown, with different subclasses. Radiation induced RMS is a rare variant of this type of sarcoma.

The prognosis of patients with PIS is poor in general, regardless of type and site, with most series reporting overall 5-year survival rates in the range of 10–30% (1-8). The prognosis appears to be related to site, reflecting the feasibility of surgical resectability. RISs of the extremities have the best prognosis, whereas those involving the vertebral column, pelvis, shoulder girdle and neck have the worst.

Although adjuvant radiotherapy and/or chemotherapy have a role in the treatment of these tumours, the extent of adjuvant radiotherapy is limited by the amount of radiation previously received, which leaves surgery as the only treatment for most of these tumours (2,3). However, using brachytherapy as an adjunctive to surgery to control residual sarcoma cells is a novel method and, to the authors’ knowledge, no study has been conducted using this modality.

This report describes a case of adult spindle-cell RMS in a patient who had previously undergone radiotherapy, treated by combined surgery and targeted radiotherapy (brachytherapy) with 5 years of follow up.
Case report

A 67-year-old Caucasian male was referred to the Head and Neck Unit, University College London Hospital (UCLH), London complaining of a painless lump in the right side of the neck which increased in size over 7 months duration. The mass did not cause any symptoms, including dysphagia or speech or breathing problems. Past medical history included hypertension and hypothyroidism. The patient is a non-smoker but chronic heavy drinker (40 units/week for over 20 years).

The patient was previously diagnosed with T3N0M0 laryngeal squamous cell carcinoma (SCC) seven years prior to his presentation to the unit. He underwent laryngectomy with radical neck dissection (level II–IV) followed by radiotherapy (66 Gy) and chemotherapy.

On clinical examination, the mass was 8×5 cm in size, and was infiltrating deep into the sternomastoid muscle. There were no palpable lymph nodes. Magnetic resonance imaging (MRI) of the neck reported a fixed solitary mass in the right neck (levels 2 and 3) arising from the sternomastoid muscle, with heterogeneous high signal intensity on T2-weighted images and low signal intensity on T1-weighted images (Figures 1 and 2). The patient had an examination under anaesthesia (EUA) that confirmed the findings, followed by an endoscopic examination of the oropharynx and larynx which was unremarkable. This was followed by an ultrasound-guided core biopsy, after which the diagnosis was confirmed histopathologically as radiation-induced high grade spindle cell sarcoma.

Agreement on the MDT was reached and the patient received single agent doxorubicin for three cycles. This was followed with extended radical neck dissection with clear margins, successfully performed; followed by reconstruction with an ipsilateral pectoralis major pedicled muscle flap and insertion of brachytherapy catheters (Figure 3). Radiation was given twice daily for three consecutive weeks. The patient had an uneventful recovery and remains disease free at his 5 year follow-up (Figure 4).

Recovery after anaesthesia was uneventful. The patient complained of discomfort in the area of surgery, with clear fluid discharge from the chest (flap donor area) for up to 2 months, which healed afterward with simple conservative dressings.

Macroscopically, a well circumscribed but not encapsulated solid, solitary red lesion was encased in thick fibromuscular tissue. Microscopic examination revealed elongated spindle-shaped cells, with vesicular nuclei, numerous mitoses and a pale cytoplasm, forming long fascicles. These cells were mixed with sparse polygonal or rounded rhabdomyoblasts with foci of sarcomeric differentiation. Immunohistological staining was positive for desmin, which confirmed the myogenic nature.
Discussion

Rhabdomyosarcoma (RMS) is a malignant mesenchymal tumour that originates from immature cells that are destined to differentiate into striated skeletal muscle. RMS predominantly affects children, constituting more than 50% of all paediatric soft-tissue sarcomas. It can be subdivided into three histological subtypes: embryonal, alveolar and pleomorphic RMS.

Spindle-cell RMS is a less common variant of embryonal RMS and is predominantly composed of spindle cells. The tumour is most often encountered in the paratesticular region in children, in whom it is generally associated with a better prognosis. The first two cases of spindle-cell RMS in adults were described by Rubin et al. 1998 (5).

Little is known about the aetiology of spindle-cell RMS in adults. In this patient, although radiotherapy was an effective treatment in the management of a malignant tumour of the larynx, radiation induced sarcoma developed. Determination of a cause-effect relationship between prior irradiation and radiation-induced tumour formation requires the following criteria: (I) documented history of irradiation in this site, (II) the new malignancy must arise within the irradiated field, (III) the new tumour must be histologically distinct from the original primary lesion, (IV) the latent period between the irradiation exposure and the development of the new malignancy must be 5 years or longer (6).

Our patient fulfilled the criteria by virtue of the tumour’s location, previous and new histology, and latency period of more than 5 years. The presenting signs and symptoms of radiation induced RMS are variable, depending on the site of initial presentation, the extent of the tumour and the presence or absence of distant metastases and lymph-node involvement. Patients generally present with a fast growing mass. Its diagnosis is difficult because of induration and fibrosis of the tissue within the former field of radiation.

Generally, radiation-induced sarcomas are associated with an outcome that is significantly poorer than that of stage-matched soft tissue and osteogenic sarcomas that arise independently of irradiation. Five-year disease-free survival rates for RIS are 10% to 30% (7). The poor prognosis of RIS in the head and neck region may be explained by the following factors: (I) delay in diagnosis, (II) proximity of the tumour to major neurovascular structures, which may place constraints on the limits of surgical resection, (III) limited treatment options because of the dangers of irradiating the previously irradiated field and the relatively poor sensitivity of these tumours to chemotherapy, (IV) RIS in the head and neck may be biologically more aggressive and (V) RIS develops from radio-resistant tumour clones and may not be responsive to further radiotherapy.

Early detection is the main hope for survival in cases of RIS because there are no preventive measures to safeguard patients with cancer against RIS apart from limiting radiotherapy or using surgery as the primary treatment modality. Thus immediate workup should be performed when any pain or
swelling occurs in an irradiated field, because the clinical diagnosis of RIS can be difficult due to fibrosis and induration within the irradiated field.

The management of radiation-induced sarcoma is challenging and differs from that of sarcoma not induced by radiation. Complete surgical excision appears to offer the best means of palliation and the only rational chance for long-term survival (8). Prognosis is related to the site and resectability; thus RIS of the extremities has a 30% 5-year cure rate whereas RIS of the vertebral column and pelvis is associated with a 5-year survival rate lower than 5%. The survival rate of RIS in the head and neck is somewhere in between these rates. In our patient, critical anatomy at the Lymph node neck levels II and III did not limit us in obtaining a wide surgical margin.

Although only limited follow-up data are available, adult spindle-cell RMS appears to have a more aggressive clinical course when compared with cases occurring in the paediatric population. Experience with treatment of adult spindle-cell RMS is limited. Treatment options include surgery, radiation therapy and chemotherapy. Patients are categorized according to their risk assessment, using a clinical group (CG) and a site-based tumour–nodes–metastasis (TNM) staging system (7).

In conclusion, despite the poor prognosis and aggressive pattern of such a rare kind of sarcoma, combining surgery with brachytherapy achieved 5 years disease free.

Consent
Written informed consent was obtained from the patient for publication of this case study and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
The authors declare that they have no competing interests.

Authors’ contributions
All authors contributed to conception and design, carried out the literature research, manuscript preparation and manuscript review. All authors have read and approved the final version of the manuscript.
References

Figures

**Figure 1:** Preoperative MRI coronal views of the radiation-induced high grade spindle cell sarcoma of the sternomastoid muscle

**Figure 2:** Preoperative MRI axial views of the radiation-induced high grade spindle cell sarcoma of the sternomastoid muscle

**Figure 3:** Surgical excision and reconstruction of the area with pectoralis major pedicled muscle flap and insertion of brachytherapy catheters

**Figure 4:** Postoperative MRI axial views of following surgery and brachytherapy