Case Report
Successful treatment of recurrent Kimura’s disease with radiotherapy: a case report

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Abstract: Kimura’s disease is a rare, chronic inflammatory disorder affecting the skin and subcutaneous tissue, predominantly in the head and neck region. It is benign but may be recurrent and difficult to eradicate. A case of recurrent Kimura disease in a 53-year-old man was reported. Radiation therapy was performed for recurrence after surgical excision twice. The prescribed radiation dose was 36 Gy. With a follow-up time of 68 months, the patient was free of the disease.

Keywords: Kimura’s disease, recurrence, radiotherapy

Introduction
Kimura’s disease (KD) or eosinophilic lymphfolliculoid granuloma is a rare benign disorder and was first described by Kim and Zeto in China in 1937 [1] and characterized by Kimura et al in 1948 [2]. The disease most commonly occurs in young adult Asian men and has a favorable prognosis. The peak age of onset is the third decade. It is a chronic inflammation lesion and predominantly involves deep subcutaneous tissue and lymph nodes of head and neck region. Kimura’s disease is often associated with regional lymphadenopathy, solitary or multiple subcutaneous nodules, sometimes with salivary gland enlargement and elevated serum immunoglobulin E (IgE) levels and peripheral blood eosinophilia [3]. Histopathology of KD shows a marked follicular hyperplasia, eosinophilic infiltrates, and proliferation of postcapillary venules [4]. Although KD is a benign condition the lesions can be very difficult to manage. Recurrence rate is very high (above 60%) after local excision [5]. Previous cases of KD treated with surgery and cyclosporine have been published. We report a patient of recurrent neck KD successfully treated with radiotherapy.

Case report
A 53-year-old Chinese man with diabetes for two years and presented with recurrence of multiple small firmed subcutaneous masses mainly on his right side neck. Two years ago, he was first found several firmed nodules on his right side neck with red and swollen skin. He was diagnosed of tuberculosis (TB) and treated with rifamycin (Nydrazid or isoniazid) and rifampin (rifampicin) for one month by local hospital and stopped treatments since the side effects of medicine on liver. However, the size of masses became larger (growing) during the treatment. After 2 months, the masses had been removed by surgery in local hospital and were diagnosed as inflammation of lymph nodes. Ten months after the surgery and nodules were recurrent at right side neck and treated by using anti-TB drugs for two months, but the treatment was terminated owing to skin allergy. Then the patient was referred to a hospital in Beijing. The masses were excision second time by surgery 10 months after recurrence. Based on the histology he was diagnosed as KD. Four months later, he developed masses third time on the same location and was referred to this hospital. Excision biopsy was performed.

Histological examination (Figure 1) revealed a reactive lymph follicular hyperplasia with prominent germinal centers that were surrounded by fibrous tissue. This was accompanied by prominent proliferative blood vessels and postcapillary venule proliferation. Numerous eosinophils infiltrated the interfollicular areas and formed
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scattered eosinophilic microabscesses in some areas. Based on these clinical and histological findings, he was made a diagnosis of recurrent KD.

Laboratory investigations revealed increased eosinophils (1.01 x 10⁹/L; normal 0.02-0.52 x 10⁹/L) and elevated serum IgE levels (537.2 IU/mL, normal 1.31-165.3 IU/mL). Results of other investigations including kidney and liver function were all within normal.

B ultrasound scan revealed that multiple small subcutaneous masses occupying the neck region. The largest one was 1.9 cm x 0.7 cm x 0.7 cm in size. CT scan showed that the largest mass was 1.7 cm x 1.1 cm x 1.1 cm (Figure 2A).

The patient was treated with X-ray radiation therapy. The prescribed radiation doses were 36 Gy, with 2 Gy per fraction, 5 fractions per week. After treatment the masses significantly reduced in size (Figure 2B). The patient’s condition has remained free from recurrence throughout the 68-month follow-up period.

Discussion

So far, the etiology for KD is still unknown [6]. Kimura’s disease is a rare benign eosinophilic folliculoid granuloma. Clinical presentation is predominantly affecting deep subcutaneous tissue and lymph nodes of head and neck region [6, 7]. Its histological characteristics are the follicular hyperplasia, eosinophilic infiltrates and proliferation of postcapillary venules [8]. The clinical and histological features of this present case are consistent with the changes of KD. Although it was developed in a common

Figure 1. Photomicrographic features of Kimura’s disease (H&E). A. Markedly reactive follicular lymphoid hyperplasia with prominent germinal centers and diffuse inflammatory cell infiltrate separated with fibrous tissue. Original magnification, x 100. B. Hyperplastic lymphoid follicle with eosinophilic infiltrate within the skeletal muscle. Original magnification, x 100. C. The interfollicular infiltration was rich in proliferative blood vessels and eosinophilic infiltrate. Original magnification, x 400. D. Eosinophils forming eosinophilic microabscesses with higher magnification. Original magnification x 400.
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location of KD, it was a rare disease and recurrent twice. So far only hundreds of patients were reported. It does not become widely recognized, and very easily to be misdiagnosed as other diseases. For current case it was misdiagnosed as TB in a local hospital and treated with anti-TB drugs. In addition to TB, differential diagnosis should also be made from subcutaneous soft tissue tumors, lymphoproliferative or granulomatous lesions, reactive lymphadenopathy, hyperplasia and benign tumors of subcutaneous vascular proliferations.

Though KD is a benign disease, its unpredictable response to the therapeutic interventions, poses a great challenge to the treating physician and the patients. There is no consensus about the optimal treatment for Kimura’s disease. The medical therapies for Kimura disease including surgical resection, regional or systemic steroid therapy, radiotherapy, cytotoxic and laser therapy have been utilized, but none has been proved to be the optimum modality [6, 9, 10]. Surgery with complete excision is difficult since the infiltrative nature of the lesion and swelling of regional lymph nodes [11]. Recurrent lesions may develop several times after initial presentation and surgical removal. It has been suggested that drug therapy is the first choice in middle-aged or elderly patients with KD [12]. Steroid therapy has been shown transiently effective, after weaning, the tumors often increase in size [4]. Recently, anti-IgE therapy has been introduced [13]. In the report there patients of KD were treated with anti-IgE (omalizumab) for four months. The size of diseased regions and the peripheral blood eosinophil were all decreased after anti-IgE therapy, but complete remission of the tumor was not observed. Radiotherapy has been used in cases of refractory disease [14], and shown to be effective for local control of KD [11, 15]. The present case provides evidence that recurrent KD can be treated successfully with radiation therapy after surgical removal. This suggests that radiation therapy may be an effective noninvasive therapeutic alternative for recurrent KD that provided local control and prevent recurrence (68 months at the time this paper was written) and without side-effects at the dose applied (36 Gy).

Disclosure of conflict of interest

None.

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Figure 2. Photographs of CT scan. A. CT scan shows one of the lesions (1.7 x 1.1 cm, arrowed) at submaxillary region, before radiotherapy. B. CT scan shows the same lesion (1.0 x 0.5 cm, arrowed), after radiotherapy.
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References


