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Prognostic factors of survival in treatment of scalp angiosarcoma in Chinese population

Sherby Suet Ying Pang*, George Kam Hop Li

Division of Plastic and Reconstructive Surgery, Department of Surgery, Queen Mary Hospital, 102 Pokfulam Road, Hong Kong, China

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ABSTRACT

Background: Scalp angiosarcoma is a rare soft tissue sarcoma with a very poor prognosis and high mortality. Due to its low incidence, prognostic factors and treatment strategies are not well defined.
Objective: This study aims to report our long-term results in terms of survival and prognostic factors affecting survival through retrospective analysis of 38 consecutive patients with scalp angiosarcoma who underwent treatment at two different plastic centers in Hong Kong.
Result: The overall survival rate for 1, 3, and 5 years of maximally treated patients was 56%, 24%, and 12%, respectively. Univariate analysis with Cox regression model showed that age >70 years (risk ratio, RR 5.0; $p = 0.02$), satellite lesion on presentation (RR 8.4, $p = 0.02$), and tumor of size >5 cm (RR 2.79, $p = 0.05$) were factors affecting the overall survival. Multivariate analysis showed that only the involved deep resection margin was a significant risk factor (RR 8.6, $p = 0.02$) However, univariate analysis showed that the involved deep resection margin (RR 10.2, $p = 0.01$) and satellite lesion on presentation (RR 0.73, $p = 0.01$) were factors affecting disease-free survival. Besides, satellite lesion on presentation predicted local recurrence on univariate analysis (RR 2, $p = 0.01$).
Conclusions: The prognosis in terms of overall survival and local disease control was poor with a 5-year survival rate of 12% in maximally treated patients.

* Corresponding author. Tel.: +852 2255 4394.
E-mail address: sherbypang@hotmail.com (S.S.Y. Pang).

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Introduction

Cutaneous angiosarcoma is a rare, aggressive soft tissue sarcoma which mainly affects elderly male.[1–5] Angiosarcomas comprise approximately 2% of all soft tissue sarcomas, with 50% being cutaneous angiosarcomas.[6] Cutaneous angiosarcomas involve the head and neck region with scalp being the most commonly affected site. They constitute only 1% of all head and neck tumors.

Cutaneous angiosarcomas most often develop spontaneously. Nevertheless, few reports on malignant transformation from preexisting vascular lesions are available.[6] They are believed to arise from endothelial cells. Radiation is a well-known risk factor for breast angiosarcomas,[7] but etiological factors for cutaneous angiosarcomas remain unclear. They usually present as a bruise or a nodule that develops into a diffuse plaque with satellite lesion rapidly.[1]

An early study in 1987 has shown that despite aggressive treatment, the median survival was around 20 months with a 5-year survival rate of 12%.[1] With the current developments in surgical techniques and adjuvant treatment, recent studies have estimated the 5-year survival rate to be 22–43%.[8–11] Patients treated with radical radiotherapy alone had a 2-year infield control rate of 67% only.[12]

Treatment of scalp angiosarcoma is a multimodality approach including radical surgery, adjuvant radiotherapy, chemotherapy, and immunotherapy. Studies show that due to the aggressive nature of the disease, prognosis is generally poor, and complicated reconstructions are fraught with frequent wound complications and delayed healing. Reasonable reconstruction options included skin graft on calvarium after the removal of the outer cortex, local flaps, and free flaps.

In this study, we had retrospectively analyzed 38 consecutive patients with scalp angiosarcoma who received treatment at two different plastic centers in Hong Kong. We report our long-term results in terms of overall survival and prognostic factors affecting survival.

Patients and methods

All patients diagnosed with scalp angiosarcoma and treated at Queen Mary Hospital from 1987 to 2014 or Kwong Wah Hospital from 2003 to 2014 were included in this study. Data were obtained by reviewing the medical records and electronic patient resources. A total of 42 consecutive patients were identified, with two being excluded due to incomplete data. Two patients were lost to follow-up. The remaining patients had either died or had up-to-date survival data and were still on follow-up at regular intervals. The resultant median follow-up of all 38 patients was 11 months (range: 1 month–12 years). Mortality was taken as the primary end point for determining overall survival. Local recurrence or distant metastasis documented in clinical records and investigations was taken as the secondary end point to determine disease-free survival.

These 38 patients with histologically confirmed scalp angiosarcoma included 32 men (80%) and six women (20%) with age range being 40–90 years (median = 77 years). A single nodular lesion over the scalp was observed in 45% of the patients (Figure 1), diffuse pattern lesion in 29%, and satellite lesion in 21% (Figure 2). The median duration between patients complaining of a scalp lesion and first seeking medical advice from primary care doctors was 6 weeks (range: 0 weeks–2 years). Patients were referred to us with a median delay of 6 weeks after their first visit to primary care doctors (range: 0 weeks–23 months). Tumor size ranged from 2 to 20 cm (median: 9 cm); 26% were T1 tumors ($\leq$5 cm) and 68% T2 tumors (>5 cm). Three patients were diagnosed with metastatic regional lymphadenopathy and two with systemic metastasis.

A total of 25 patients (66%) were treated with radical surgery, eight (21%) with palliative radiotherapy to the scalp, and two (5%) with palliative chemoirradiation or immunotherapy, while three patients (8%) refused treatment. In the case of the 25 patients who underwent wide local excision of the scalp, the radial resection margins were 1–5 cm (median: 3 cm); craniectomy was performed in five patients by neurosurgeons for deep margin control. Wide excision resulted in a scalp defect of largest diameter (range: 7–30 cm, median: 16 cm). Six defects (24%) were reconstructed by skin grafting, four (16%) by local flaps, six (24%) by muscle-only free latissimus dorsi flap along with skin grafting, and six (24%) by free anterolateral thigh myocutaneous flap (Figure 3). Seventy-six percent patients received adjuvant radiotherapy to the scalp; four patients underwent chemotherapy, while one underwent both chemotherapy and immunotherapy.