內科學誌 2005 ：16 ：146-150

Rhabdomyosarcoma of the Adult Nasopharynx ： A Case Report

Yueh-Lan Huang, Chin-Feng Tseng, Li-King Yang, and Chen-Hua Tsai

Division of Oncology, Department of Internal Medicine,
Cardinal Tien Hospital

Abstract

Rhabdomyosarcoma is a rare and highly malignant neoplasm of the adult head and neck with a significant incidence of metastases. The outcome for adults with this disease is poorly documented.

Despite its ominously poor prognosis, combined polychemotherapy, radiotherapy and surgery, whenever feasible, has markedly improved survival rates in some patients. We report the case of a 67-year-old man who presented with a three-week history of a rapidly growing mass on the right upper neck with no symptoms of nasal obstruction, pain in the ears, tinnitus, or headache. Magnetic resonance image (MRI) study of the head and neck revealed a mass in the right and posterior walls of the nasopharynx with extension to the right parapharyngeal space, sphenoid and ethmoid sinuses and enlarged bilateral cervical lymph nodes extending from the submandibular to the supraclaricular regions. Nasopharyngeal biopsy showed poorly differentiated small, blue round cells. A diagnosis of rhabdomyosarcoma was confirmed by positive staining for myo D1, myogenin, desmin and CD56. Combination chemotherapy with VAC (vincristine, adriamycin and cyclophosphamide) regimen and radiotherapy was completed within 5 months. Follow up MRI of the head and neck, bone scan and chest X-ray 18 months later revealed neither local recurrence nor distant metastasis. (J Intern Med Taiwan 2005; 16: 146-150)

Key Words ： Rhabdomyosarcoma, Adult nasopharynx, Radiotherapy, Chemotherapy

Introduction

Soft tissue sarcomas account for 15-20% of all pediatric malignancies, of which rhabdomyosarcoma is the most common. Rhabdomyosarcoma may be defined as a malignant tumor of the rhabdomyoblasts with a microscopic picture simulating that of striated muscle cells. In children and young adults, rhabdomyosarcoma tends to occur in the head and neck, extremities, and genitourinary tract[1]. In contrast, rhabdomyosarcoma rarely occurs in the adult head and neck but commonly presents as truncal and extremity tumors. The relative proportion of pleomorphic tumors also increases with age. Rhabdomyosarcoma is classified into two major histologic subtypes, namely, juvenile (embryonal and alveolar) and adult

Correspondence and requests for reprints : Dr. Chen-Hua Tsai
Address : 362, Chung Cheng Road, Hsintien City, Taipei County, Taiwan 23137

pleomorphic[2]. Due to nonspecific initial manifestations, diagnosis is often made late in its course. Metastatic disease most commonly occurs in the lungs, bone marrow, bones, liver and brain, and poor response to chemotherapy are strongly associated with worse prognosis[3]. However, combined chemotherapy and radiotherapy have considerably improved survival in recent years[4].

Case report

A 67-year-old man presented with a three-week history of a rapidly growing, non-tender mass in the right cervical region. Upon consultation with an otorhinolaryngologist, he was told to have nasopharyngeal cancer with cervical lymph node metastases. Nasopharyngeal biopsy revealed poorly-differentiated small blue round cells but no definitive diagnosis was given. He was subsequently referred to our Oncology clinic for further evaluation and management.

Physical examination revealed the presence of two confluent fixed, firm non-tender masses, one on each side of both upper cervical regions (7x7x5 cm on the right side and 5x6.5x4 cm on the left side, respectively), and a protruding mass in the right nasopharyngeal cavity. However, the patient was aymptomatic and denied having symptoms of nasal obstruction, pain in the ears, tinnitus, nor headache. Magnetic resonance image (MRI) study of the head and neck showed a mass in the right and posterior walls of the nasopharynx with extension to the right parapharyngeal space and enlarged bilateral cervical lymph nodes extending from the submandibular to the supraclavicular regions (Fig.1).

A second biopsy revealed a tumor with small to medium-sized blue round cells, many of which had eccentric nuclei and eosinophilic cytoplasm and some tumor cells with scanty cytoplasmic substance. Postive immunohistochemical staining for desmin, myo D1, myogenin, CD56 and actin was compatible with a diagnosis of rhabdomyosarcoma (Fig. 2). The

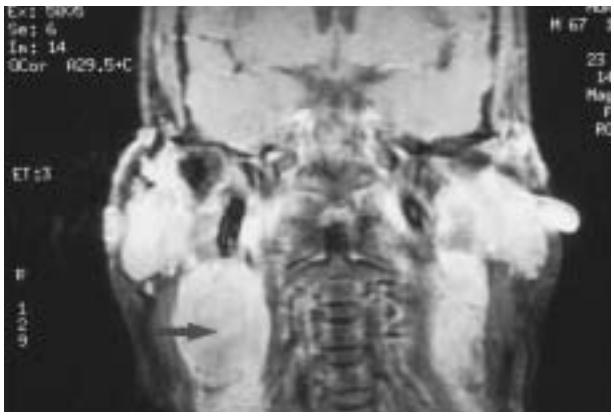

Fig. 1. Axial and coronal section T1-weighted MRI scan showing a mass (arrow), 7x7x5 cm in the right and posterior wall of the nasopharynx with extension to the right parapharyngeal space with enlarged bilateral cervical lymph nodes from the submandibular to the supraclavicular regions.

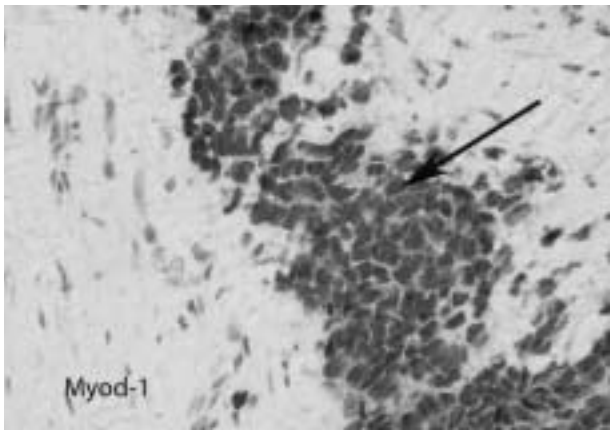

Fig. 2. The tumor cells stained positive for myoD1, myogenin, CD56 which were compatible with a diagnosis of rhabdomyosarcoma. (Immunohistochemical staining x 200)

cytokeratin stain was negative. Systemic work-up including bone scan, bone marrow biopsy, chest X-ray and abdominal sonography revealed no metastatic lesion. Three courses of chemotherapy with vincristine ($1.4mg/m^2$), adriamycin ($50mg/m^2$), cyclophosphamide ($750mg/m^2$) and cisplatin ($70mg/m^2$) every 4 weeks were given with granulocyte colony-stimulatory factor (G-CSF) 5mcg/kg/d support. However, the patient still developed one episode of severe febrile neutropenia (absolute neutrophil count less than 50/cmm) after the second course of chemotherapy. Radiotherapy (4500cGy in 180cGy/fx) and biweekly vincristine ($1.0mg/m^2$) were given for maintenance therapy. The entire course of treatment

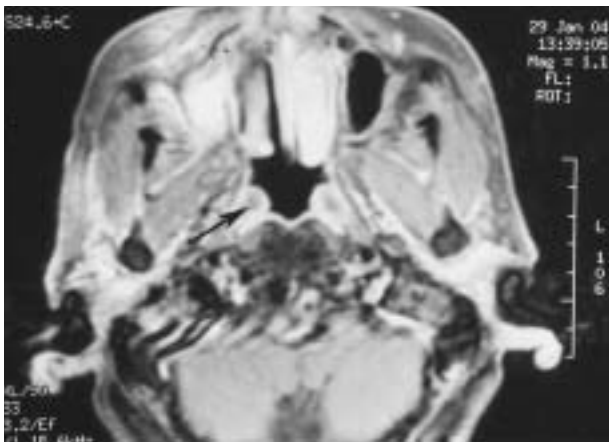

Fig. 3. Eighteen months after therapy, MRI of the neck and nasopharynx revealed neither local recurrence nor distant metastasis.

was completed within 5 months. Follow-up MRI of the head and neck (Fig. 3), bone scan, and chest X-ray done 18 months later revealed neither local recurrence nor distant metastasis.

Discussion

Rhabdomyosarcoma, a tumor derived from mesenchymal tissue, was first described by Weber in 1854[2]. It is the most common malignant tumor in children but is quite rare in adult [5]. Thus, current literature regarding rhabdomyosarcoma is mostly derived from pediatric research studies .This tumor can develop anywhere due to its myogenic nature but occurs mostly in the head and neck, genitourinary tract, and extremities of children. In adults, the proximal part of the extremities and the genitourinary tract are the most common sites[1]. There are four histologic variants, namely, embryonal, alveolar, pleomorphic and undifferentiated. However, some authorities have proposed that there are only two major variants, that is, the juvenile (embryonal and alveolar) and the adult pleomorphic forms. Unfortunately, accurate histologic classification was limited by insufficient biopsy specimen in this patient.

Sarcomas develop from connective tissues such as muscles, fat, and membranes lining joints or blood vessels. However, nasopharyngeal carcinoma usually originates from cells lining the oropharynx or that part of the pharynx which lies behind the oral cavity.

Patients with nasopharynx cancer may notice a lump in the neck or develop pain or ringing in the ears. Patients may also complain of impaired hearing, frequent headaches. However, the primary lesion in head and neck rhabdomyosacroma usually presents as a non-tender mass or localized swelling with no accompanying nasopharyngeal symptoms, which is how our patient presented.

Since histopathologic finding in this patient revealed poorly differentiated small, blue round cells, differential diagnosis had to include extraskeletal Ewing's tumor, primitive neuroectodermal tumor, neuroblastoma, malignant lymphoma, or intra-abdominal desmoplastic small round cell tumor. However, positive immunohistochemical staining for desmin, myo d1, myogenein, CD56 and actin is compatible with a diagnosis of rhabdomyosarcoma in our patient.

In nasopharyngeal rhabdomyosacroma, the lungs and bones are the usual sites of distant metastasis. Newman and Rice reported distant metastases in 50% of their patients after treatment, 62% of whom had local recurrence prior to the development of metastasis. Moreover, 90% of distant metastases occurred 2 years after treatment, and the 5-year survival rate of patients with distant metastases was a dismal 0%[6]. Staging study in our patient showed tumor extension to the right parapharyngeal space, with enlarged bilateral neck lymph nodes from the submandibular area to the supraclavicular regions.

Management of rhabdomyosarcoma usually involves a combination of surgery, radiotherapy and chemotherapy. Total tumor resection confers the most favorable treatment outcome. However, complete tumor resection is often difficult because of its invasive nature of the tumor and its often anatomically canceled location[7]. Overall prognosis for rhabdomyosarcoma has improved since the introduction of multidisciplinary treatment[1] but prognosis remains poor for patients with metastatic disease at presentation and

poor response to chemotherapy[8]. Radiotherapy is targeted at the pre-chemotherapy sites of disease. Chemoradiation following complete surgical resection appears to be the ideal therapy[9]. Since the adult head and neck rhabdomyosarcoma is biologically different from tumors occurring in children, there is little evidence to support any particular therapeutic program[4]. The optimal treatment remains undefined and multimodal approaches combining surgical resection, radiation, and systemic chemotherapy need further investigation.

In conclusion, adult nasopharyngeal rhabdomyosarcoma rarely occurs but should be kept in mind, nevertheless, the differential diagnosis of a head or neck mass and biopsy finding of poorly differentiated small blue round cells.

Despite its poor outcome and severe complications, adequate local control of adult rhabdomyosarcoma may be attainable with multidisciplinary treatment and proper supportive care.

References

1.Atsuo Tsuchiya, Hiroshi Furukawa, Yoshiyuki Endo, et al. Rhabdomyosarcoma presenting as an anterior cervical mass in an adult: report of a case. Surg Today 2000; 30: 1107-9.

2.Kanagalingam J, Medcalf, M, Courtauld E, et al. Rhabdomyosarcoma of the adult nasopharynx. ORL J Otorhinolaryngol 2002; 64: 233-6.

3.Esnaola NF, Rubin BP, Baldini EH, et al. Response to chemotherapy and predictors of survival in adult rhabdomyosarcoma. Ann Surg 2001; 234: 215-23.

4.Wight RG, Harris, Shortland JR, et al. Rhabdomyosarcoma of the nasopharynx: a case with recurrence of tumor after 20 years. J Laryngol Otol 1988; 102: 1182-4.

5.Huseyin, Sercan, Arzu, et al. Laryngeal involvement of rhabdomyosarcoma in an adult: case report. World J Surg Oncol 2003; 1: 17.

6.Newman AN, Rice DH. Rhabdomyosarcoma of the head and neck. Laryngoscope 1984; 94: 234-9.

7.Weng TH, Wang TZ, Tsai ST. Nasopharyngeal rhabdomyosacroma-case report. J Taiwan Otolaryngol Head Neck Surg 2001; 36: 48-52.

8.Ward IG, Blackstein M, Catton C, et al. Rhabdomyosarcoma of the head and neck: patterns of relapse. Oral presentation, 9th Internastional Congress on Oral Cancer 2003; 16-19.

9.Flamant F, Rodary C, Rey A. SIOP MMT 84 study: results on rhabdomyosarcoma (RMS) with 4 years median follow-up (abstract). International Society of Paediatric Oncology, SIOP XXIIIth Meeting, 1991; 434-5. Abstract.

成人鼻咽横紋肌肉瘤

黃月蘭　曾金鳳　楊麗瓊　蔡振華

天主教耕莘醫院　內科部腫瘤科

摘　要

成人頭頸部腫瘤中，橫紋肌瘤是罕見且具高轉移率的極惡性腫瘤，有關其存活率的文獻報告也並不多。儘管其預後極差，但有報告指出藉由化學治療、放射治療及外科手術等多科合作治療方式，有某些病人仍可提高存活率。在此報告一病例。一位六十七歲男性病人於三週前發現右上側頸部快速明顯變粗，但無鼻塞、耳痛、耳鳴或頭痛現象。頭頸部的核磁共振攝影顯示腫瘤由鼻咽右後壁延伸至右側咽喉部，並同時發現由下顎延伸至上鎖骨之雙側頸部淋巴亦已腫大。鼻咽部位的切片呈現出小深藍圓形分化不良之癌細胞，此並非一般鼻咽癌細胞。且此細胞對myoD1 、myogenin 、desmin 及CD56 等特殊染色方式呈陽性反應，因此確認此爲少見之橫紋肌肉瘤。病人接受爲期五個月的積極化學治療 (VAC vincristine, adriamycin and cyclophosphamide) 及放射治療，在第十八個月後追蹤 (包括頭頸部核磁共振攝影、骨頭掃描、胸部X 光)，並無局部復發或遠處轉移的現象。