

Synovial sarcoma of the parapharyngeal space

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Abstract

Synovial sarcoma is a rare soft tissue sarcoma in the head and neck region and parapharyngeal space. A 21-year-old girl presented with a 6-month history of progressive right arm pain, neck mass and upper aerodigestive tract obstruction. On physical examination there was a large painless mass arising from the right-sided parapharyngeal space causing airway obstruction and with no cervical lymphadenopathy. Initial magnetic resonance imaging (MRI) revealed a large tumor in the right-sided parapharyngeal space. She underwent near total resection of the tumor. Pathologic report disclosed the diagnosis of synovial sarcoma. She then received postoperative adjuvant external radiotherapy to the primary site and a dose of 60 Gy was delivered. Less than 8 months after the completion of the treatment she developed widespread lung metastases. Herein we describe the clinical, radiological and pathological finding of the case.

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1. Introduction

Sarcomas of the head and neck region are a rare and diverse group of neoplasms and only account for less than 1% of all neoplasms that occur in this area [1]. Synovial sarcoma usually occurs in young adults and is found in the paraarticular areas of the tendon sheaths and joints in the lower- and upper-extremity. This soft tissue tumor accounts only for 3–10% of all head and neck soft tissue sarcomas [2,3]. Neoplasms arising from the parapharyngeal space are uncommon and account for only 0.5% of all the head and neck region. Salivary gland and neurogenic tumors are the most common neoplasms involving the parapharyngeal space [4]. Herein we describe a case of parapharyngeal synovial sarcoma.

2. Case presentation

A 21-year-old girl with no significant past medical history, presented with a 6-month history of progressive right arm pain, followed by a 3-month history of right-sided otalgia and jaw and shoulder pain, dysphagia, dyspnea and cervical mass. On physical examination, she had a large painless mass arising from the right-sided parapharyngeal space causing remarkable airway narrowing with no cervical lymphadenopathy. Initial magnetic resonance imaging (MRI) revealed a large contrast-enhanced mass arising from the right-sided parapharyngeal space causing airway obstruction and pressure effect on the major cervical vessels (Fig. 1). The patient underwent near total tumor resection and a fragile spongy, non-encapsulated 4 cm × 3 cm × 2 cm mass was resected. Histopathological examination showed a hypercellular tumoral tissue with biphasic pattern (Fig. 2). The epithelial component composed of glandular structures lined by cuboidal cells (Fig. 3). The stromal component composed

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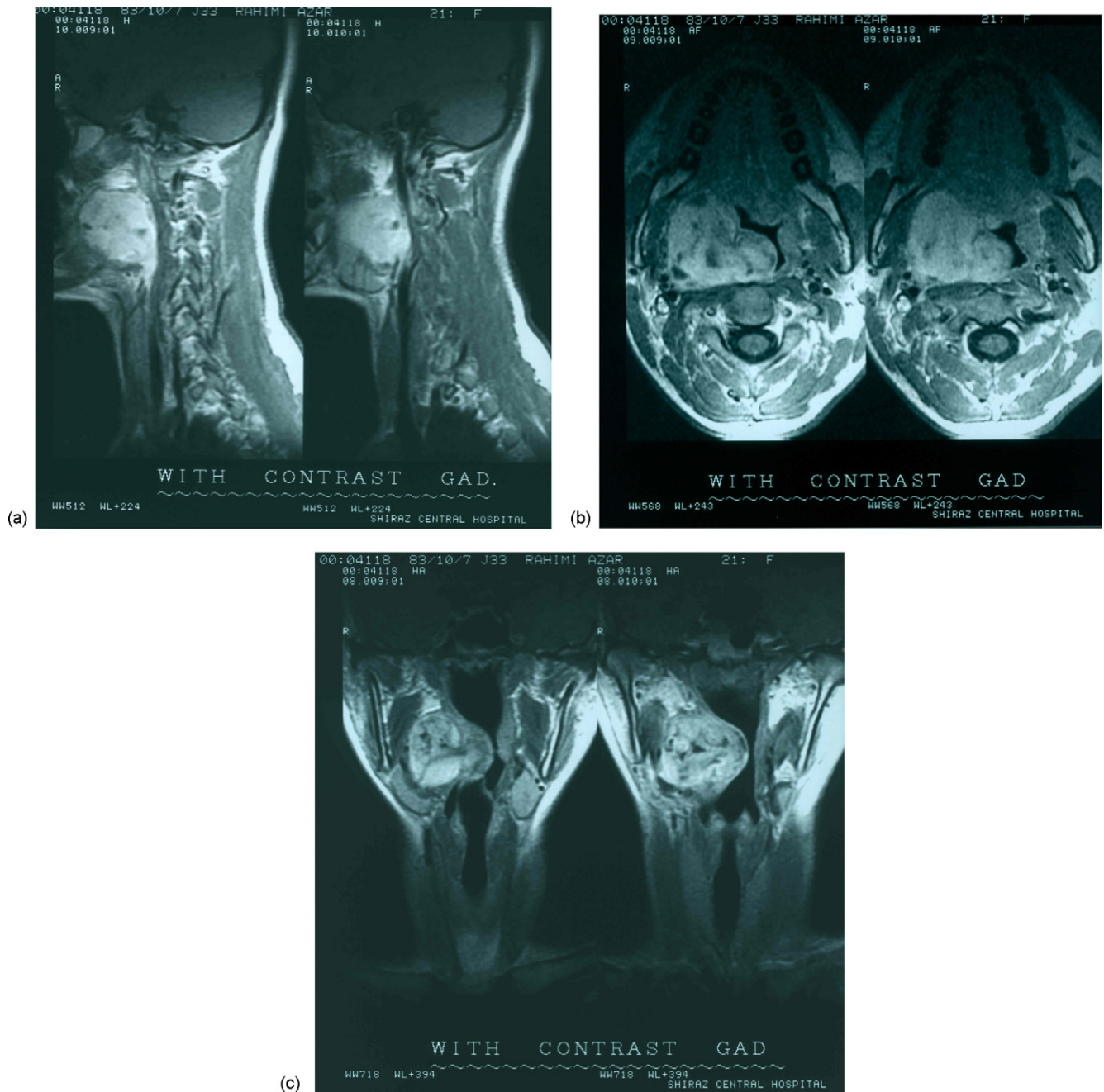


Fig. 1. (a–c) Sagittal, axial and coronal contrast-enhanced MR images of the right parapharyngeal synovial sarcoma.

of oval to spindle cell arranged as sheet and fascicles (Fig. 4). Immunohistochemical study revealed cytokeratin and EMA positively in the epithelial component and diffuse Vimetin positivity in spindle cells (Fig. 5a–c). Postoperative chemoradiation was considered as adjuvant therapy. External beam radiation therapy was performed using 9 MV X-rays from a linear accelerator, with daily fraction of 2 Gy, 5 fractions per week and a total dose of 60 Gy was delivered. The spinal cord was excluded from the radiation fields after 45 Gy. Concomitant weekly cisplatin (30 mg/m^2) as radiosensitizer was combined with the radiation therapy. In less than 8 months after the completion of multi-modality therapy, she

developed multiple lung metastases. She developed two local recurrences during 18-month follow up.

3. Discussion

Parapharyngeal space, a potential fascial plane of the head and neck, may be involved by the neoplastic disease which represents less than 1% of all head and neck tumors. Eighty percent of parapharyngeal neoplasms are benign and 20% are malignant [5]. Benign salivary gland and neurogenic tumors are the most common neoplasms

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