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ALVEOLAR RHABDOMYOSARCOMA OF THE HAND: A VERY RARE CASE OF SOFT TISSUE SARCOMA

Elde Alveolar Rabdomyosarkom: Yumuşak Doku Sarkomlarinda Oldukça Nadir Bir Olgu

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ABSTRACT

Soft tissue sarcomas of extremities are very rare and generally have a bad prognosis. Rhabdomyosarcomas are soft tissue sarcomas that, while extremely rare in adults, are one of the most common neoplasms in children and adolescents. Extremities are unusual sites of origin for rhabdomyosarcomas and usually associated with unfavorable outcome. Complete surgical removal of the tumor represents a major factor in terms of survival.

Our study presents the case of primary alveolar rhabdomyosarcoma of the left hand in a 22 years old male who was treated by fifth ray amputation after an incomplete tumor excision.

The patient is still tumor-free after 4 years following operation.

Key Words: Hand; hypothenar; tumor; sarcoma; rhabdomyosarcoma; amputation.

ÖZET

Ekstremitelerin yumuşak doku sarkomları çok nadir ve oldukça kötü bir prognoza sahiptir. Rabdomyosarkomlar yumuşak doku sarkomu olup, erişkin yaşta nadir olarak görülürler, daha sıklıkla çocukluk ve ergenlik dönemi tümörleridir. Ekstremite tutulumu rabdomyosarkomlar için alışılmadık bölgeler olup ve genellikle kötü sonuçlarla ilişkilidir. Sağ kalım açısından major faktör tümörün cerrahi olarak tamamen çıkarılmasıdır.

Vakamız 22 yaşında erkek bir hastanın sol elinde çıkan primer alveolar rabdomyosarkomunun inkomplet tümör eksizyonu sonrasında 5. ray ampütasyon ile tedavisini anlatılmaktadır.

Hastanın ameliyat sonrası 4 yıllık takiplerinde tümör dokusu saptanmamıştır.

Anahtar Kelimeler: El; hipotenar; tumor; sarkom; rabdomyosarkom; amputasyon.

INTRODUCTION

Rhabdomyosarcoma (RMS), which has an exceedingly poor prognosis, is the most common malignant soft tissue sarcoma of muscle origin in children and adolescents (1, 2).

Three histological types occur: embryonic, alveolar and pleomorphic. The embryonic type is the most common and typically occurs in children, the alveolar type tends to affect older children and young adults and frequently is found in the extremities (2). The unfavorable prognosis group includes alveolar RMS with 54% of 5 year survival. Anatomic sizes, tumor size, nodal and distant metastases, adequacy of tumor resection are the factors that influence the prognosis (2, 3).

Tumors arising in the extremities generally have a worse prognosis. Higher percentage of these tumors is alveolar RMS (2,3). Although RMS of extremities are usually small in size, they often present with disseminated disease at initial diagnosis (3,4). Histological subtype is often alveolar and has been generally associated with a poorer prognosis than that of other subtypes (2-4).

CASE REPORT

A 22 years old male patient was admitted to The Hospital with a 5 months history of painless, non-mobile, hard mass on the left hypothenar eminence, which had slowly been increasing in size for the past three months. There was no history of injury, fever or weight loss. Clinical examination revealed a non-mobile and non-tender mass on the hypothenar eminence. The overlying skin was not inflamed. The range of motion of the wrist and fingers were within normal limits. There was no neurovascular disturbance.

A soft tissue shadow was seen in the hypothenar space on plain radiographs, while MRI showed

a lobulated mass measuring 5x4x3 cm without infiltration of bone with low signal intensity on T1 weighted images and heterogenous high signal intensity on T2 weighted images. There was a cystic necrotic component at the superior part of the lesion. Adductor digiti minimi muscle and musclotendinous junction were surrounded by the mass (Figure 1).

An excisional biopsy was performed. Macroscopically, excised mass measured 5x4x3 cm in dimension. Grossly, the tumor was gray-tan and rubbery in consistency. Cut surface was solid with focal microcystic change. Histologically, the tumor was composed of groups of small, oval cells with scant cytoplasm seperated and surrounded by fibrous septa. The cells at the periphery of the groups were adhered to fibrous tissue, whereas the cells at the center were loosely arranged. The tumor was infiltrating the muscle tissue and entrapment of normal muscle fibers was detected, both histologically and immunohistochemically (Figure 2). Occasional multinucleated giant cells were also present within the tumor. Immunohistochemical studies revealed diffuse positive staining with vimentin and desmin in the tumor cells.

Because of positive surgical margins, the patient underwent fifth ray amputation. Post operatively the patient received external beam radiotherapy (RT) with a dose of 6 cycles, as well as chemotherapy, consisting of doxorubicin / izofosfamide. He returned to his work at 3 months postoperatively. 6 months after fifth ray amputation, the patient presented with sudden onset of pain over the ulnar side of the fourth metacarpal. Local recurrence was investigated by control MRI and a lesion in 1.5x1 cm dimensions with focal destruction of the neighboring fourth metacarpal was seen (Figure 3). The patient rested for one month. Afterwards complete resolution which confirms overuse of the extremity was seen on the MRI (Figure 4). Abdominal and thorax CT findings were in normal limits. There was no metastatic lesion. Four years after the operation the patient had ability to use his hand in daily life.

DISCUSSION

RMS is one of the most typical tumors of childhood, accounting for more than 50% of soft tissue sarcomas (1, 5, 6).

It is a very aggressive tumor that tends to invade contiguous structures as well as becoming disseminated via the lymphatics and blood stream (1, 2, 5, 6).

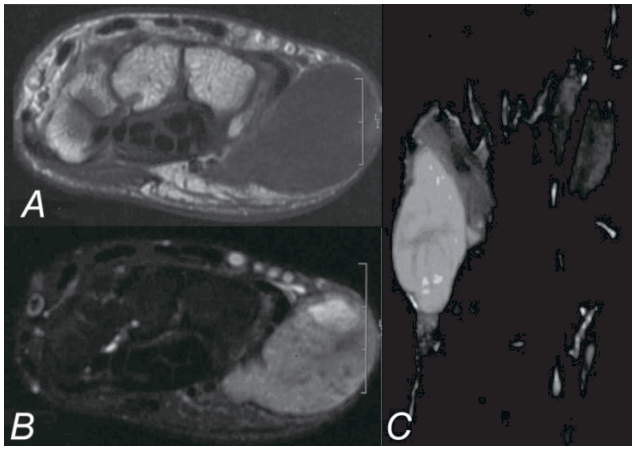

Figure 1: A mass lesion with lobulated contours located at the hypothenar part of the left hand with low signal intensity on T1 weighted images and heterogeneous high signal intensity on T2 weighted images. Both cortical and medullar signal intensity of fifth metacarpal are within normal limits, thus MRI findings are compatible with soft tissue malign mass lesion.
A: Axial T1 weighted, B: Fat suppressed Axial T2 weighted, C: Coronal fat suppressed T2 weighted.

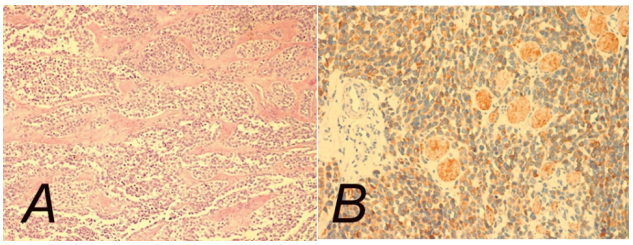

Figure 2:
A: Diffusely infiltrating tumor cells loosely attached to fibrous septa peripherally with central loss of cohesion (H&E x100).
B: Diffuse desmin positivity within the tumor cells and the entrapped normal muscle fibers (Immunohistochemistry x 200).

About 15% of all RMS arise in the extremities and they usually have a worse outcome than those occurring elsewhere (5-7). RMS of the extremity has been reported as more aggressive, in recently published median survival rates varying from 30 months to 55 months (6-8). Limb RMS is reportedly associated with the unfavorable alveolar subtype with a greater frequency of nodal involvement. Although there have been few reported cases of hand RMS, involvement of the hand is extremely rare (2, 5-8).

The most common sites for metastasis from primary RMS are lung, bone, bone morrow and liver (5). There is a predilection for the reproductive organs and breast predominantly in female patients. All tumors metastasizing to these organs were interestingly alveolar RMS (2, 5).

Many patients have metastatic disease in lymph nodes that are clinically normal (5, 9). Lymph node metastases, however significantly worsen the prognosis. RT has been used selectively in order to enhance local tumor control (2, 5, 9).

Limb sparing by wide local excision of the tumor is the treatment of choice but amputation may occasionally be necessary (2, 7, 9,). However, patients who were treated primarily with amputation paradoxically had lower survival rates than those who had primary wide, local excision (6-10). Therefore, primary amputation should keep in mind in the cases of failed wide excision and advanced local RMS as in our case (1, 2, 5-10).

Many of the soft tissue sarcomas about the hand occur painlessly, avoiding diagnosis for months to years. In most patients, wide local surgical resection with adjuvant therapy is the treatment of choice for RMS (9, 10). There is a need of more data to clarify best treatment modality.

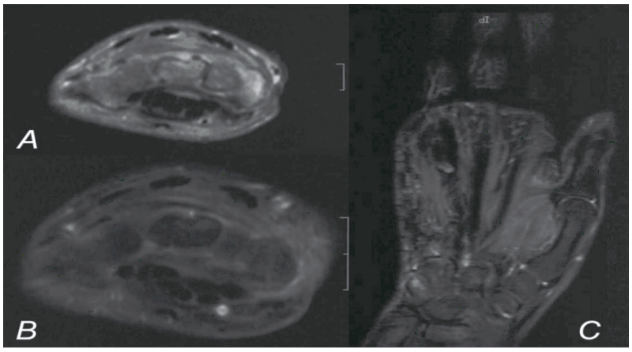

Figure 3: Early postoperative MRI. A lesion in 1.5x1cm dimensions which is hiperintense on T2 weighted, hypointense on T1 weighted images with contrast enhancement. Decreased medullar signal on T1 weighted and increased signal on T2 weighted images with focal destruction of the neighboring fourth metacarpal.
A: Axial post contrast T1 weighted, B: Axial fat suppressed T2 weighted, C: Coronal fat suppressed T2 weighted.

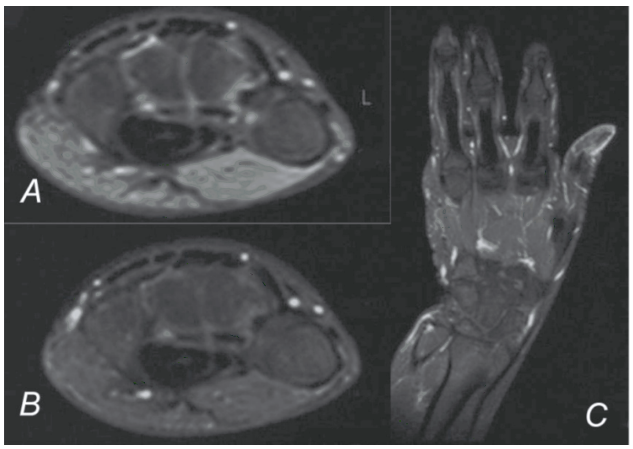

Figure 4: Control MRI after one year. The lesion and focal destruction of the fourth metacarpal that was seen on the early postoperative MRI was disappeared. No residual tumor is seen.
A: Axial fat suppressed T2 weighted, B: Axial fat suppressed T1 weighted, C: Coronal fat suppressed T1 weighted.

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