Göğüs Ön Duvarında Radyoterapi Sonrası Geç Dönemde Gelişmiş Bir Dev Anjiyosarkom Olgusunun Tanısal Görüntüleme Değerlendirimi

DIAGNOSTIC IMAGING EVALUATIONS OF A POSTIRRADIATION GIANT ANGIOSARCOMA LOCALIZED ON CHEST ANTERIOR WALL ARISED IN LATE PERIOD

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Özet

Radyoterapi sonrası sarkomlar sık görülmemesine rağmen, değişik tipte malign tümörler için uygulanan radyoterapiyi takiben gelişen angiosarkomlar bildirilmiştir. Bu çalışmada cerrahi olarak total ekstirpasyonunu gerçekleştirdiğimiz ve etiyolojisinde 13 yıl önce Hodgkin hastalığı nedeniyle gördüğü radyoterapi sonrası geç dönemde geliştiğine inandığımız 30 yaşındaki bir hastada saptanmış sağ ön hemitoraks üst kadranındaki dev angiosarkom olgusunu tanı ve cerrahi tedavi yönünden literatür ışığı altında inceledik.

Anahtar kelimeler: Angiosarkom, radyoterapi sonrası, malign vasküler oluşumlar

Türk Göğüs Kalp Damar Cer Derg 2003;11:255-257

Summary

Although sarcomas are occasional after irradiation, there are angiosarcomas developed after radiotherapy for different type malign tumors. In this study we evaluated the imaging methods performed for diagnosis of a giant angiosarcoma developed at right anterior hemithorax upper quadrant, in late period after radiotherapy in a 30 years old patient irradiated 13 years before for Hodgkin disease and in whom we totally extirpated the angiosarcoma surgically.

Keywords: Angiosarcoma, postirradiation, vascular tumors

Turkish J Thorac Cardiovasc Surg 2003;11:255-257

Introduction

Angiosarcomas are malignant vascular tumors [1]. They frequently occurred in subcutaneous tissues of skin and breast and deep soft tissues [2]. Angiosarcoma is one of the rarest form of soft tissue neoplasm and it was shown in the 20 year study held in M.D. Anderson Hospital that they have an insignificant rate in total vascular tumors and they comprise less than 1% of all sarcomas [3].

Case Report

The patient was a 30 years old man and admitted to our clinic for a big, enlarging and aching mass at right upper quadrant of chest anterior wall, for the last 3 months. We determined that he was treated with combined chemotherapy and radiotherapy for a year, 13 years before, for Hodgkin lymphoma diagnosis. We inspected and palped a nonpulstatil, solid, 10x15 cm mass on right upper quadrant of chest anterior wall.

Diganostic imaging investigations were withheld respectively and in chest X-ray we observed a homogenous, 10x7 cm mass between 2^{nd} and 5^{th} ribs, extending convexly to right lung,

adjacent to mediastinum at right hemithroax upper zone. Following this investigation we performed a soft-tissue ultrasonography directed to relevant localization and found a 10x12 cm mass, arising from right superior-anterior mediastinum and extending subcutaneously, having a heterogenous solid inner structure with cystic and calcific areas and an appearance on sternum probably due to destruction and prodiagnosed as thymoma (?) or malign teratoma (?) (Figure 1). The next method was contrasted thoracic MRI and we determined a space-occupying lesion localized at right clavicle distal end's cauda and thorax anterior wall (a suspicious infiltration at clavicle's distal end) (Figure 2). Lesion was at medial line and at right of it. Its largest axial dimension was 10x8cm. Craniocaudal extension was approximately 8cm. It was showing heterogenic signal changes in all sequences and a heterogenic type strong uptake pattern (Figure 2). It was partially invaded the clavicle's distal part from right caudal region. It deplaced the pectoral muscle to anterior in the right (infiltration in right pectoralis majus muscle). Also it was showed a close neigbouring with right brachiocephalic artery and vein and brachiocephalic truncus. It was extending to distal part of ascending aorta at caudally in anterior mediastinum and



Figure 1. Heterogenous solid mass with cystic and calcific areas at soft tissue ultrasonography.

very adjacent to vascular structures. Right 1st rib was in the mass and had an cranial indentation at 2nd rib. (Figure 3). Last imaging investigation was aortography and it showed that there wasn't contrast diffusion in right upper hemithoracal mass.

During operation, mass invased the 1^{st} and 2^{nd} ribs, _ medial of clavicle, 1/3 lateral part of manubrium sterni and sternoclavicular joint and destruction was significant in these structures. We completed the radical excision of the mass. Also destructed anterior segments of 1^{st} and 2^{nd} ribs and anterior _ medial part of clavicle were removed with excision. Open defect due to removal of pleura adjacent to mass was repaired secondarily with 4x6 cm goretex patch (Figure 4). Skin defect at right infraclavicular region reconstructed with left pectoral myocutaneous flap closure. Patient was discharged on the 8th



Figure 3. In this section, 1^{st} rib is in the mass and mass indentates the 2^{nd} rib cranially.



Figure 2. Contrasted thoracic MRI showed an suspicious infiltration at distal end of clavicula on front wall of thorax.

postoperative day with surgical cure. In the interpretation of hystopathologic investigation it was determined to be an postirradiation angiosarcoma due to Hodgkin disease therapy and that bone tissues were infiltrated (moderately differantiated, grade II).

Discussion

Angiosarcomas are malignant tumors that recapitulate many of the functional and morphologic features of normal endothelium [1,3]. In the past it was shown that intraabdominal or abdominal wall angiosarcomas have occurred following radiotherapy for cervix, ovary or uterus carcinomas. Same event occurred at different localisations after radiotherapy for



Figure 4. Secondary repair of open defect due to removal of pleura adjacent to mass with gore-tex patch.

different malign or benign conditions [3].

Sarcoma occurrence at radiotherapy region for breast carcinoma or various malign neck tumors is rare. However they can be arise at bone or soft tissues. This type of tumors are announced so infrequently. Interval between radiotherapy and occurrence of sarcoma is very variable and its generally over 10 years but rarely can be seen in 5 years (4). In another study, the latent period between radiotherapy and diagnosis of vascular lesion was determined between 3 and 20 years [5]. Growing pattern of an epithelioid angiosarcoma is very similar to the carcinoma showing radiation changes [4].

When literatures published after 1963, were rewieved Kardamakis [6] reported 20 cases with only mediastinal angiosarcoma. Tumor mostly localized at anterior mediastinum. Therapy was surgical excision and prognosis was bad. Value of the adjuvant chemotherapy not fully understood yet, because so few cases reported.

Key of successful therapy in chest wall tumors is early diagnosis and agressive surgical resection [4,6]. Diagnostic aproach in a patient with a probable chest wall tumor must contain; carefull history taking, physical examination, laboratory assays and following conventional chest radiography, soft tissue USG, computerized chest tomography [1]. Also magnetic resonance imaging (MRI) is a preferential method for primary chest-wall tumor. MRI not only distinguishes tumor, from nerve and blood vessels but also helps imaging from different planes as coronal and sagital, so we preferred this method too. Superiority of CT, can be seen during evaluation of lung parancim's metastatic diseases.

Preferred therapy method in angiosarcoma cases is extended resection of the lesion [3,5]. Radiotherapy and/or chemotherapy can be used as adjuvant therapy.

In the recent years there are developments in diagnosis and therapy of angiosarcoma and the most important one is vascular endothelial growth factor (VEGF) as announced by Fujimoto and assoc. [7]. VEGF is an angiogenic cytokine and it is used in tumor angiogenesis study during recent years. While stimulating growth, VEGF also increases the permeability of endothelial cells. Some angiosarcomas can produce both VEGF and its receptors. This investigation showed that angiosarcoma can grow with both autocrine and paracrine ways. Although we can't monitorize the angiosarcoma therapy with serial VEGF serum concentration measures practically, hope full studies are going on.

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