Granulocytic Sarcoma Presenting as Epidural Mass with Cord Compression in Aleukemic Patient

Alösemik Hastada Omurilik Basısı Yapan Epidural Granülositik Sarkoma

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Abstract: Granulocytic sarcoma (GS), constituted by the immature cells of myeloid serial, is a rare, solid tumor. The tumor may be observed in the course of systemic myeloid leukemia. Occasionally, granulocytic sarcoma may be observed without findings of acute leukemia. Authors report a 17-year-old female who had had spinal cord compression due to GS without evidence of leukemia. Up today, only 6 cases have been reported with paraparesis due to spinal epidural granulocytic sarcoma without clinical symptomatology of leukemia. Laminectomy is mandatory for cases of cord compression and radiotherapy must be performed after the surgical intervention. For cases without spinal cord compression, chemotherapy and radiotherapy may be used together.

Key Words: Chloroma, granulocytic sarcoma, spinal cord compression

Özet: Myeloid serinin immatür hücrelerinden gelişen granülositik sarkom (GS), nadir solid tümördür. Tümör, sistemik myeloid löseminin seyri esnasında görülebilir. Nadiren granülositik sarkom akut löseminin bulgusu olmaksızın görülebilir. Yazarlar, lösemi bulgusu olmaksızın GS' a bağlı spinal kord kompresyonu olan 17 yaşındaki bir kadın olgu bildirmiştir. Günümüze kadar, löseminin klinik semptomatolojisi olmaksızın spinal epidural granülositik sarkoma bağlı paraparezi gelişen sadece 6 olgu bildirilmiştir. Kord kompresyonu olan olgularda laminektomi zorunludur ve radioterapi cerrahi girişimden sonra uygulanmalıdır. Spinal kord kompresyonu olmayan olgularda kemoterapi ve radioterapi birlikte kullanılabilir.

Anahtar Sözcükler: Kloroma, granülositik sarkom, omurilik basısı

INTRODUCTION

Granulocytic sarcoma (GS), constituted by the immature cells of myeloid serial, is a rare and solid tumor (8, 9, 12, 14, 17). Usually it occurs in young patients with acute myelocytic leukemia (9, 13, 14). Owing to its greenish color caused by the myeloperoxydase enzyme in the tumor cells, it is also referred to as chloroma (7, 11, 13, 17).

The most common tumor sites are the skin, the lymph nodes, the chest, the abdomen, the soft tissues, the bones, the sinuses, and the orbita, but the central nervous system is a rare location (4, 5, 6, 9, 11, 13-15). Occasionally, granulocytic sarcoma may be observed without findings of acute leukemia (2, 5, 8, 10-14, 16, 18, 20). So far, only 6 cases have been reported who had paraparesis due to spinal epidural granulocytic sarcoma without clinical symptomatology of leukemia (2, 10, 11, 16, 18 ,20).

We report a case of granulocytic sarcoma that caused cord compression without any findings of leukemia.

CASE REPORT

A 17-year-old female was referred to us for evaluation of weakness in the legs that had been progressing for 5 days. Two months earlier, she had developed a low-back pain. One week before the admission, the pain had radiated into both legs.

Neurological examination revealed weakness in both legs, with the right side being more affected than the left side. Hypoesthesia was observed on the right below the level L1 and on the left below the level L2. There was loss of deep tendon reflexes in both legs.

All laboratory tests were within the normal range.

Roentgenograms of the lumbar and thoracic spine showed no pathology. A myelogram showed complete obstruction at the L1- L2 level (Figure 1). Myelo-CT showed a hypodense, extradural mass at the L1-L2 level (Figure 2).

The patient underwent laminectomy from Thoracic 12 to Lumbar 2. A fragile green tumoral mass, located extradurally, had enveloped the cord and invaded the laminae and paravertebral regions. Subtotal resection was performed.

Histopathological examination revealed diffuse infiltration which was heterogenous without a remarkable organization in the soft tissue. Cells forming the infiltration, polymorphic, pleomorphic in shape, with partially broad eosinophylic cytoplasm, atypical cells (with no similarity to each other) could be seen. There were polymorphonuclear leucocytes in between. The immunohistochemical study showed lysozyme, LCA positive, CD 15 focal and weak positive in the tumor cells. Histopathological diagnosis was confirmed as granulocytic sarcoma (Figure 3). Further examinations of the blood and the bone marrow for leukemia were negative.

The postoperative course was uneventful, and the patient was referred to an oncology clinic for radiotherapy. For a period of 34 months of follow up, the patient was free of any neurological symptoms, and normal radiological findings were observed.

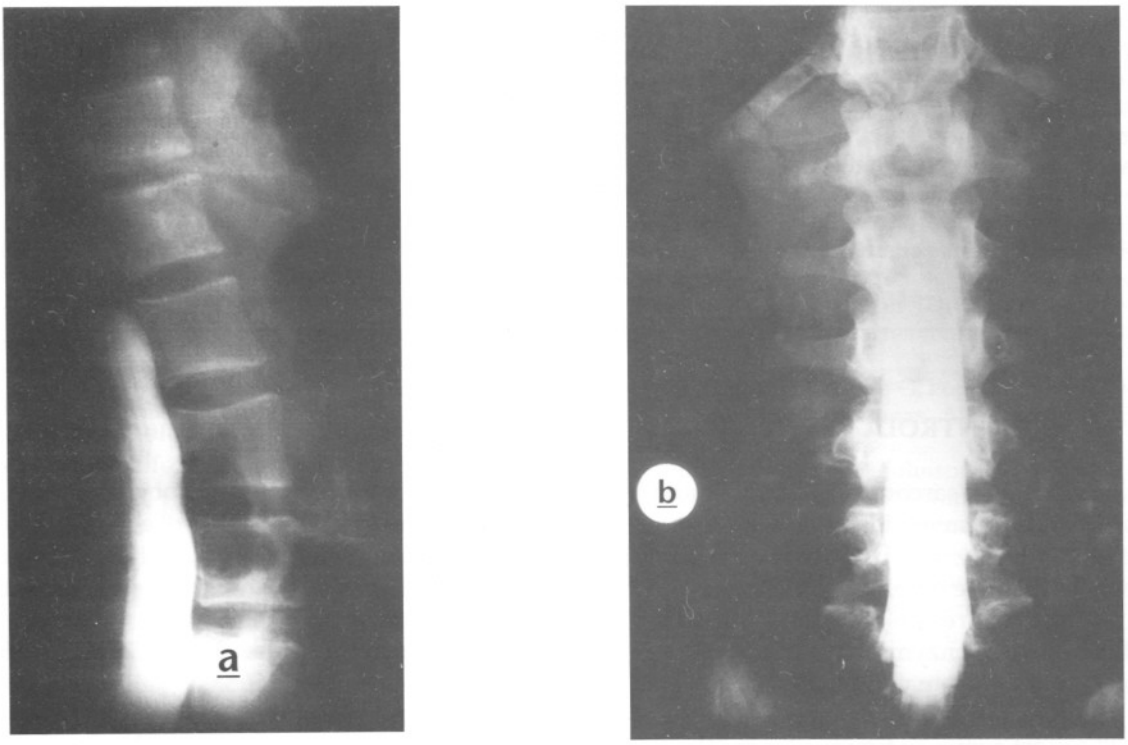

Figure 1, a and b: Myelography showing complete obstruction at the L1-L2 level.

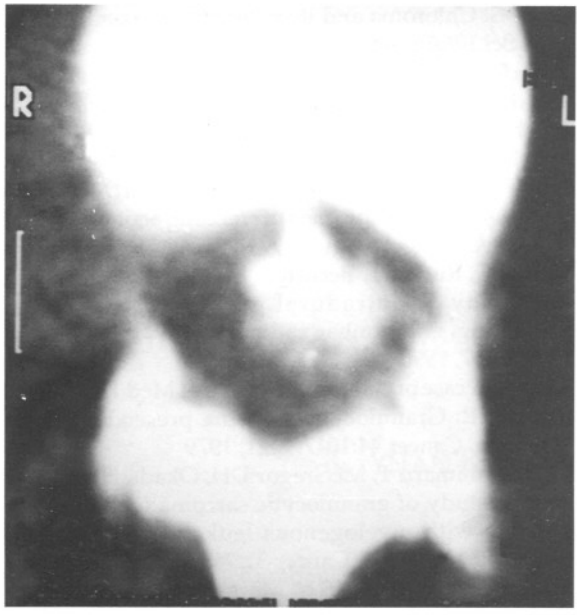

Figure 2: CT-myelography showing the extradural mass compressing the dural sac.

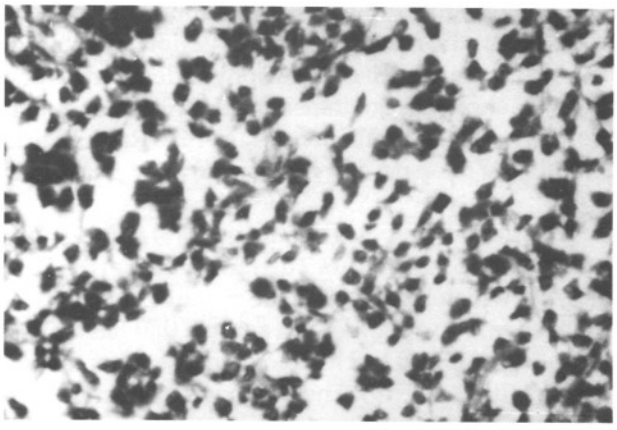

Figure 3: Photomicrograph of the tumor demonstrating diffuse infiltration which is heterogenous without a remarkable organization in the soft tissue (H&E x 200).

DISCUSSION

Granulocytic sarcoma is a rare, solid tumor of the immature cells of myeloid serial (8, 9, 12, 14, 17). It was first described by Burns (1) in 1811, in a patient with proptosis and a green retroorbital tumor. The term " chloroma " was given to the lesion by King (7) in 1853, because these tumors often display a greenish color. In 1893, Dock (3) established the association between chloroma and leukemia. In the following years, the terms of chloromyeloma, chloromyelosarcoma, granulocytic leukosarcoma, myeloblastoma, myelosarcoma, and myelocytoma were used. Rappaport (17) used the term of granulocytic sarcoma in 1966, which has been widely used since then.

Granulocytic sarcoma is usually observed in the course of acute leukemia but it is also occasionally seen in the course of chronic leukemia or myeloproliferative disorders (9, 13, 14). The incidence of granulocytic sarcoma in acute myelogenous leukemia has been reported as 2.9 %, but it has been reported as high as 3-8 % in the autopsy series (8, 9, 13). Granulocytic sarcoma occurs more commonly in children, as 60 % of the patients with granulocytic sarcomas are under 15 years of age (9). Our case was a 17- year-old female.

Although the tumor may occur in any location, the most common sites are the skin, the lymph nodes, the chest, the abdomen, the soft tissues, the bones, the sinuses, and the orbita, but the central nervous system location is rare (8, 9, 12, 14, 17). Occasionally, it may occur when the blood and bone marrow findings are absent (2 , 5, 8, 10-14, 16, 18, 20). Numerous cases of spinal cord compression secondary to granulocytic sarcoma have been reported (9, 13-15, 21), but these have all been in association with leukemia. In literature, there were only six cases in which granulocytic sarcoma was the cause of cord compression without systemic findings of acute myeloid leukemia (2, 10, 11, 16, 18, 20) . In our case preoperatively or postoperatively, no findings of leukemia were present.

Although Neiman et al. (14) report approximately 10.5 months for the findings of leukemia to become apparent in cases with aleukemic granulocytic sarcoma, Meis et al. (12) report 4 patients with no evidence of leukemia 3,5 to 16 years after diagnosis. In our case, this interval has reached 34 months.

Most of the spinal lesions occured as extradural masses (15, 18, 20). Thoracic region of the spine is most frequently involved (15, 20). Back pain and radicular pain were present in more than 60 % of the reported cases and preceded neurological deficit by a median of 4 weeks (20). Once symptoms suggesting spinal cord compression began, progression was often rapid (15).

In radiological studies of the spine,

roentgenograms show bone destruction, myelograms show partial or complete obstruction, CT shows contrast-enhancing isodense or hypodense mass and the MRI shows hypointense mass on T1 images and isointense mass on T2 images (4, 15) . Radiological findings of our case correlates well with literature.

Although pathological diagnosis is easy because of the green color due to the myeloperoxidase enzyme which brings forth the name of chloroma, it is not as easy to diagnose in a sample that has waited for a long time and lost its color because of air contact (5, 11, 9, 10-14, 17, 19). The diagnosis of granulocytic sarcoma can be confirmed by electron microscopy, by the napthol AS-D chloroacetate esterase stain, and by immunoperoxidase staining with antilysozyme (8, 10-12, 14, 17, 19). Histologically, it is constituted by poorly differentiated immature cells (8, 10-14, 19).

In the differential diagnosis, lymphoma, rhabdomyosarcoma, amelanotic melanoma, and metastatic should be considered (8, 12, 18).

In the treatment, laminectomy is mandatory for cases of cord compression and radiotherapy must be performed after the surgical intervention (11, 13, 15, 18, 20, 21). If a systemic illness is present, chemotherapy should be added to the therapy regiment (18, 21). Although radiotherapy alone is effective, chemotherapy alone is not effective (21). For cases without spinal cord compression, chemotherapy and radiotherapy may be used together (21).

We conclude that in leukemic cases with spinal cord compression, granulocytic sarcoma should be considered. In addition, follow-up is mandatory in aleukemic cases with epidural granulocytic sarcoma due to the fact that frank leukemia may occur.

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