Mediastinal Hemangiopericytoma with Bone Metastases in a 2.5 Year-old-boy

Kemik Metastazı olan Mediastinal Hemanjioperisitomalı 2.5 Yaşında Bir Erkek Olgu

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ABSTRACT

Hemangiopericytoma (HPC) is a soft tissue sarcoma characterized by the proliferation of capillary pericytes. Here we present a 2.5 year-old-boy with a mediastinal mass and multiple bone metastases. The tumor was very aggressive and showed progression despite aggressive chemotherapy, leading to a dismal prognosis.

Key Words: Rare tumors, bone metastases, pediatric oncology

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ÖZET

Hemanjioperisitoma, kapiller perisitlerin proliferasyonu ile karakterize bir yumuşak doku sarkomudur. Burada, mediastinal bir kitle ve birden çok kemik metastazları olan 2,5 yaşında bir erkek olgu sunulmaktadır. Tümör çok agresif seyretmiş ve yoğun kemoterapiye ragmen ilerleme göstererek kötü bir prognoza yol açmıştır.

Anahtar Sözcükler: Nadir tümörler, kemik metastazı, pediatrik onkoloji

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INTRODUCTION

Hemangiopericytoma (HPC) is a malign mesenchymal tumor characterized by the proliferation of capillary pericytes (1,2). It accounts for less than 3% of all soft-tissue sarcomas in children older than 1 year of age (3). Mediastinal HPC is extremely rare and few data are available about the management and prognosis of these patients. Here we present a case with a mediastinal mass and widespread bone metastases.

CASE REPORT

Routine echocardiographic examination of a 2.5-year-old boy operated for a ventricular septal defect at the age of 6 months revealed a mass with smooth borders behind the right atrium, between the right pulmonary artery and aorta. The physical examination revealed a pansystolic murmur of 3/6 grade at all cardiac regions and a chest scar due to the operation. The chest X-ray and the computerized tomography of the thorax showed a homogeneous, hypodense solid mass of 6×6×7 cm in the right mediastinum,

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surrounding subaortic branches of the aorta, and compressing cardiac chambers (Figure 1a,b). A bony destruction with both lytic and sclerotic metastases was detected in distal metaphyseal regions of long bones (Figure 1a). The patient underwent surgery with a median sternotomy approach, but only a biopsy could be obtained from the highly hemorrhagic mass which invaded the peripheral tissue. The histopathologic examination showed malignant cells varying from spherical to elliptic in shape with fine chromatin and no nucleoli. Tumor cells were stained focally positive with smooth muscle actin and desmin, but negative with CD1A, S-100 and myogenin. Blood vessels were stained positively with CD34. Two or three mitotic figures were observed per power field (10x) (Figure 2a,b). The tumor was classified as T2b, N0, M1 and Group IV according to Tumor-Node-Metastasis (TNM) and Intergroup Rhabdomyosarcoma Study grouping systems, respectively. Adjuvant chemotherapy consisted of various combinations of carboplatin, epirubicin, vincristine, actinomycin-D, ifosfamide, and etoposide. As there was no regression in the primary mass and bone metastases following three courses of chemotherapy, salvage chemotherapy with ifosfamide 1800 mg/m^2/day for 5 days, carboplatin 400mg/m^2/day for 2 days, and etoposide 100 mg/m^2/day for 5 days for each course was started. No radiotherapy was administered because of the potential severe cardiac and pulmonary toxicity. However, a further progression of the primary tumor and bone metastases were observed following four courses of chemotherapy. The patient who developed recurrent pathologic bone fractures at lower extremities was withdrawn from the follow-up by his family.



Figure 1 (A) Lytic, sclerotic, destructed diffuse bone metastases and solid mass in the right mediastinum (arrows) **(B)** Homogeneous, hypodense solid mass with a diameter of 6×6×7 cm in the right mediastinum, pressing the cardiac chambers (tumor labeled with arrows).

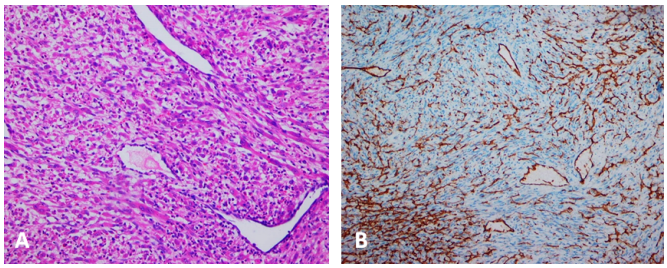


Figure 2 (A) Branched vascular structures with hematoxylin and eosin stain in the power field (20x) **(B)** Blood vessels stained positively with CD34 in the power field (20x)

DISCUSSION

Hemangiopericytoma is mainly seen in adulthood. Only 5–10% of HPC occur in children and up to 40% of patients present during the first year of life, thus considered to be congenital. The so-called adult-type was generally accepted to occur in children older than 1 year-old. Immunhistochemically, these tumors are positive with vimentin and show no epithelial, neural, and myogenic differentiation (1-4). The Pediatric Oncology Group grading system defines infantile HPCs as tumors occurring during the first four years of life. The age distinction is directly related with prognosis as infantile/congenital HPC has a more benign course with an excellent response to chemotherapy and even occasional spontaneous regression, while adult type HPC is usually aggressive with a malignant behavior (4). Our 2.5 year-old-patient showed a dismal prognosis in concordance with an adult-type HPC.

Childhood HPC is most frequently localized at the head and the neck, lower extremities, and the retroperitoneal region but it can occur in any part of the body. The pulmonary localization is considerably rare and infiltration to chest wall and pleura may cause cough, hemoptysis, and dyspnea (5-9). Interestingly, our patient didn't suffer from respiratory problems and was diagnosed while being evaluated for the cardiac disease. Johnson et al. reported a 3 year-old-girl with primary pulmonary HPC in 1992 who presented with an obstructive airway disease. This was the first child over 1 year of age reported to have an HPC localized to the lung, and was treated with surgery only (6). Subsequently, two consecutive case reports with 4 and 5 years of age by Rafaat et al. and Simonton et al. emphasized the poor prognosis of these tumors due to anatomic site, local invasion leading to recurrences, and tendency to bleeding (7,8). Recently, Horikawa-Kyo et a.l reported a 3 year-old-girl with mediastinal HPC successfully treated with surgery, radiotherapy and chemotherapy despite positive resection margins (9). To our knowledge, our patient is the fifth mediastinal HPC in children after the first year of life and the unique case with bone metastases presented in the English literature. He has a similar age with the previous cases but was presented with extensive bone metastases and showed disease progression after chemotherapy.

The most important prognostic factors in HPC are achieving local control and the presence of metastasis. Although difficult to perform because of local invasion and high vascular content of the tumor, complete resection is recommended and repeated surgical excisions may be required with localized tumors (8-10). Only a biopsy could have been conducted in our patient as the tumor had invaded peripheral tissues and would easily bleed during surgery. Efficiency of adjuvant radiotherapy and chemotherapy are also controversial (2,3). In our patient, we couldn't obtain a remission even with an intensive chemotherapy and the disease progressed despite an aggressive treatment. On the other hand, radiotherapy has been used in the case of incomplete or marginal surgical excision (10). However, no radiotherapy was administered to our patient in order to not to cause a severe cardiac and pulmonary toxicity.

CONCLUSION

The reported data about mediastinal HPC in children are limited. Our patient is one of the few cases with childhood HPC who was older than 1 year of age with mediastinal primary and unique to have bone metastases. Our case confirms the aggressive nature of HPC after the first year of life. Non-responsiveness to chemotherapy suggests that efficient treatment methods are needed for such uncommon aggressive malignancies.

Conflict of interest
No conflict of interest was declared by the authors

REFERENCES

1. Stout AP, Murray MR. Hemangiopericytoma: a vascular tumor featuring Zimmerman's pericytes. Ann Surg 1942;116:26-33.
2. Ferigo N, Cottalorda J, Allard D, et al. Successful treatment via chemotherapy and surgical resection of a femoral hemangiopericytoma with pulmonary metastasis. J Pediatr Hematol Oncol 2006:28:237-40.
3. Rodriguez-Galindo C, Ramsey K, Jenkins JJ, et al. Hemangiopericytoma in children and infants. Cancer 2000; 88:198-204.
4. Ferrari A, Casanova M, Bisogno G, et al. Hemangiopericytoma in pediatric ages: a report from the Italian and German Soft Tissue Sarcoma Cooperative Group. Cancer 2001;:92:2692-8.
5. Yousem SA, Hochholzer L. Primary pulmonary hemangiopericytoma. Cancer 1987;59:549-55.
6. Johnson JA 3rd, Warren ET, Heath BJ. Primary pulmonary hemangiopericytoma in a 3-year-old child. Tex Heart Inst J 1992;19:146-7.
7. Raafat F, Cameron AH, Mann JR, et al. Recurrent hemangiopericytoma of the chest wall: report of a case in a 5-year-old boy. Pediatr Pathol 1994;14:19-25.
8. Simonton SC, Swanson PE, Watterson J, et al. Primary mediastinal hemangiopericytoma with fatal outcome in a child. Arch Pathol Lab Med 1995;119:839-41.
9. Horikawa-Kyo Y, Tanaka T, Tanano H, et al. Mediastinal hemangiopericytoma. Pediatr Blood Cancer 2009;53:206-7.
10. Atkinson JB, Mahour GH, Isaacs H Jr, et al. Hemangiopericytoma in infants and children. A report of six patients. Am J Surg 1984;148:372-4.