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Case Report

Pediatric Spinal Cord Myeloid Sarcoma Presenting as Neuroblastoma: Case Report

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Abstract

A 5-year-old boy, presented a paraparesis and was referred for pelvic neuroblastoma based on MRI finding. The BMA performed in this context revealed an AML M2 with t(8;21) confirmed. The diagnosis of Myeloid sarcoma was confirmed by the histological study of the mass biopsy. The treatment initiated urgently as neuroblastoma was switched to AML protocol. The complete remission was achieved after induction 1 but the patient died from sepsis after consolidation 3.

In conclusion, MS should be considered as diagnosis in children presenting with motor impairment and spinal mass. The diagnosis can be confirmed by BMA but if the bone marrow is not involved the histological and immunohistochemistry studies are mandatory using the adequate markers. Complete remission can be achieved using chemotherapy and steroids with a good prognosis. In Morocco it is necessary to improve supportive care as well as survival in patients with leukemia.

ABBREVIATIONS

MS: Myeloid Sarcoma; AML: Acute Myeloid Leukemia; BMA: Bone Marrow Aspirate; CR: Complete Remission; CNS: Central Nervous System; MRI: Magnetic Resonance Imaging

INTRODUCTION

Myeloid sarcoma (MS) or chloroma is an extramedullary tumor composed of immature myeloid cells most commonly found in association with acute myeloid leukemia (AML) [1]. The chloroma may be the initial manifestation of leukemia and can occur in different anatomic sites. MS is most commonly found in the orbit or periorbital and the diagnosis can be difficult especially when they precede the bone marrow involvement [2]. About 47% of MS are initially misdiagnosed mostly as malignant lymphoma but also as neuroblastoma, rhabdomyosarcoma, Ewing sarcoma or peripheral neuroectodermal tumor [3]. In the CCG report 11% of patient presented a MS and 1% of them had a central nervous system CNS-MS (eg brain, spinal cord). Patients presenting CNS-MS had a significantly better survival than patients with non-CNS MS, with CSF leukemia, or with no extramedullary leukemia [4].

We report the case of a 5-year-old boy patient with a spinal cord MS referred and treated initially as a neuroblastoma.

CASE REPORT

A 5-year-old boy, with no past medical history, consulted a general pediatrician for 3-month history of lower limb pain. Recently he suffered from motor disturbance with mild urinary frequency and incontinence. Magnetic resonance imaging (MRI) revealed a thoracic and pelvic laterovertebral mass with spinal canal extension and the patient was referred to our unit with the diagnosis of neuroblastoma.

Clinical examination at the admission revealed a right peripheral facial paralysis, paraparesis predominant in the right lower extremity without tumor syndrome or evidence of bone marrow involvement. Computerized tomograpgy and MRI of the spine confirmed the double location, thoracic and pelvic, of a latero-vertebral solid mass with extension inside the vertebral canal T5-T7 and from L5 to the coccyx. Cerebral MRI was normal (Figure 1). White blood cell count was 5800 /µl without blasts, hemoglobin level was 7.7 g/dl and platelet count was 89 000/ µl. Urine catecholamine were negative.

Intravenous Methylprednisolone (2 mg/kg) associated to neuroblastoma chemotherapy (vincristine, cyclophosphamide, adriamycine) was initiated at day 1 of admission. The patient underwent simultaneously an echo guided biopsy of the mass and

a bone marrow aspiration (BMA). At day 3 the diagnosis of AML 2 was established with a BM infiltrated at 65% of blasts 100% myeloperoxydase (MPO) positives, presence of Auer rods and azurophilic granules. Cytogenetic study showed a translocation (8; 21).

Histological study of the mass showed atypical neoplastic cells with proliferation of small round hyperchromatic nuclei. Immunohistological study confirmed the diagnosis of MS (CD33 and myeloperoxydase positive) (Figure 2).

The chemotherapy was switched to Moroccan AML protocol and the patient received 2 induction of low dose aracytine associated with adramycine and 3 consolidation with high dose aracytine associated with asparaginase. Complete remission (CR) was achieved after induction 1 (improvement of motor impairment, BMA without blasts, absence of MS). After consolidation 3 the patient presented a severe neutropenic fever and died from sepsis.

DISCUSSION

A dozen pediatric spinal MS cases similar to ours have been reported in the literature. It is predominant in male and the thoracic spine is the most common site (64%) followed by the lumbar (29%), sacral (20%) then cervical (5%) spine [3]. The diagnosis is usually difficult especially before the manifestation of AML. In the present case, the patient was managed as neuroblastoma based on MRI finding and BMA was performed in this context. The diagnosis of chloroma and AML was easier in this case because of the infiltration of the BM.

Understanding MRI characteristics of this disease may help us making differentiate diagnosis. MS appears as soft-tissue mass with homogeneous signal intensity, marked enhancement and absence of cystic change, calcification and necrosis. When the imaging features above are present in sacral region without a definite leukemia history, MS could be strongly suggested especially if defused infiltration signal of bone marrow is present simultaneously. Clinicians should be alerted for timely bone marrow and peripheral blood examinations to prevent incorrect or missed diagnosis [5].

Morocco is a low/middle income country LMIC with limited resources and where pediatric oncology is facing challenges typical in similar countries [6]. Thus it is necessary to optimize how we use our resources. In the present case more importance should have been given to clinical symptoms (facial paralysis) to avoid unnecessary and expensive workup (biopsy, urine cathecholamine).

The histomorphological diagnosis of MS can be challenging to pathologists, especially in the absence of a known hematological disorder. Differentiation of granulocytic sarcoma from malignant lymphomas and other small round cell tumors is very critical [7]. A minimal panel of immunohistochemical markers should

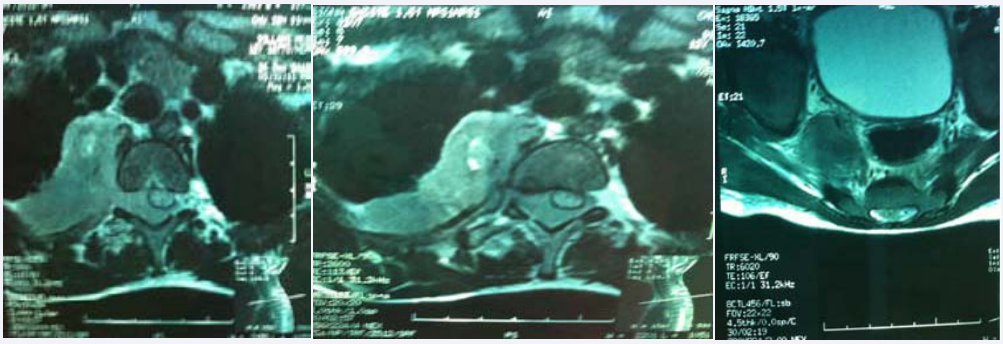

Figure 1 Lumbar MRI showing a paravertebral soft tissue mass with extension in the vertebral canal L5 –S5. The mass is in intermediate signal T2 and hyposignal in T1 enhanced on post contrast T1. Absence of calcification, peripheral capsule or necrosis.

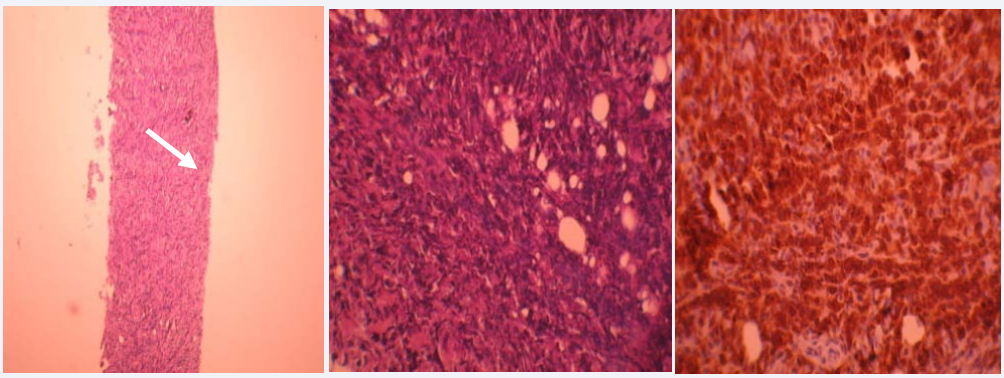

Figure 2 Histological study of the mass showed atypical neoplastic cells with proliferation of small round hyperchromatic nuclei. Immunohistological study confirmed the diagnosis of MS (CD33 and myeloperoxydase positive).

include anti-CD43 or anti-lysozyme as a lack of immunoreactivity for either of these sensitive markers would be inconsistent with a diagnosis of myeloid sarcoma. Use of more specific markers of myeloid disease, such as CD33, myeloperoxidase, CD34 and CD117 is necessary to establish the diagnosis [8]. In our case, the biopsy of the mass was unnecessary because the BM study was sufficient for establishing the diagnosis of AML M2 with t(8;21).

In the COG study a higher incidence of M2 morphology and t(8;21) was reported in patients with CNS MS. Patients with orbital MS and CNS MS had a better survival than those with non-CNS MS (92% and 73% vs 38%, $p<0.001$), as well as a better survival than patients with CNS leukemia and patients with neither MS nor CNS leukemia. The significantly higher incidence of t(8;21) in the orbital MS and CNS MS patients was not the explanation for the superior outcome. The recent COG de novo AML protocol AAML0531 did not recommend the use of radiation therapy to treat patients with MS unless the mass was causing a significant deficit [4]. In a small study of pediatric MS, none of the 15 patients received local radiation therapy and these patients had a better disease free survival compared to patients with AML and no MS [9].

The treatment is based on intensive timing chemotherapy and high dose of steroids should be added in case of motor deficiency. The surgery is not necessary to achieve a CR as reported in the present case where CR was obtained under steroids and chemotherapy.

Unfortunately, the patient died from sepsis during an episode of neutropenic fever. In recent report about infection in children with leukemia in Morocco, the median number of febrile episodes in AML was three per patient, and the rate of deaths per febrile illness was 11.3%. This higher rate of infectious deaths in leukemia compared to that reported in high-income countries, suggests that improvements in infection care and prevention, including consistent access to rapid hospitalization, diagnostics and antibiotics; and standardizing quality of patient care are necessary to improve as well as survival in patients with leukemia in Morocco.

In conclusion, MS should be considered as diagnosis in children presenting with motor impairment and spinal mass. Understanding MRI characteristics may help in making differentiate diagnosis. The diagnosis can be confirmed by BMA but if the bone marrow is not involved the histological and immunohistochemistry studies are mandatory using the adequate markers. Complete remission can be achieved using chemotherapy and steroids with a good prognosis. In Morocco it is necessary to improve supportive care as well as survival in patients with leukemia.

REFERENCES

1. Vardiman JW, Thiele J, Arber DA, Brunning RD, Borowitz MJ, Porwit A, et al. The 2008 revision of the World Health Organization (WHO) classification of myeloid neoplasms and acute leukemia: rationale and important changes. Blood. 2009; 114: 937-951.
2. Brown LM, Daeschner C 3rd, Timms J, Crow W. Granulocytic sarcoma in childhood acute myelogenous leukemia. Pediatr Neurol. 1989; 5: 173-178.
3. Landis DM, Aboulafia DM. Granulocytic sarcoma: an unusual complication of aleukemic myeloid leukemia causing spinal cord compression. A case report and literature review. Leuk Lymphoma. 2003; 44: 1753-1760.
4. Johnston DL, Alonzo TA, Gerbing RB, Lange BJ, Woods WG. Superior outcome of pediatric acute myeloid leukemia patients with orbital and CNS myeloid sarcoma: a report from the Children's Oncology Group. Pediatr Blood Cancer. 2012; 58: 519-524.
5. Anqi X, Siqing H, Zhenlin L, Chao Y. Sacral canal myeloid sarcoma as initial manifestation of granulocytic leukemia: MRI features and differential diagnosis (with a case report). Turk Neurosurg. 2014; 24: 281-283.
6. Arora RS, Eden T, Pizer B. The problem of treatment abandonment in children from developing countries with cancer. Pediatr Blood Cancer. 2007; 49: 941-946.
7. Markoc F, Bozdogan N, Yükrük FA, Gumuc EB, Akdur NC. Granulocytic sarcomas: difficulties in diagnosis. Tumori. 2010; 96: 149-153.
8. Klco JM, Welch JS, Nguyen TT, Hurley MY, Kreisel FH, Hassan A, et al. State of the art in myeloid sarcoma. Int J Lab Hematol. 2011; 33: 555-565.
9. Schwyzer R, Sherman GG, Cohn RJ, Poole JE, Willem P. Granulocytic sarcoma in children with acute myeloblastic leukemia and t(8;21). Med Pediatr Oncol. 1998; 31: 144-149.
10. Depasse J, Caniza MA, Quessar A, Khattab M, Hessissen L, Ribeiro R, et al. Infections in hospitalized children and young adults with acute leukemia in Morocco. Pediatr Blood Cancer. 2013; 60: 916-922.

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