

A case of plasmacytoma of scalp in pediatric thalassemia human immunodeficiency virus positive child

Swapan Kumar Mallick, Debashis Chatterjee, Dipanjan Majumder, Krishnangshu Bhanja Choudhury¹

Departments of Radiotherapy, Calcutta Medical College and Hospital and ¹RG Kar Medical College and Hospital, Kolkata, India

ABSTRACT

A 9-year-old female child, known case of beta thalassemia major and human immunodeficiency virus seropositive, presented with two slowly growing masses in the temporoparietal region of the skull on both sides. Irregular bony erosion at left parietal area with underlying scalp hematoma with similar change at the right side to a lesser degree were found. Fine needle aspiration cytology from masses showed abundant plasmacytoid cells some of which are binucleated along with free erythroblasts. Cytological features were suggestive of plasmacytoma. Serum protein electrophoresis showed no M-band. Urine was negative for Bence-Jones protein. Bone marrow biopsy showed no plasma cell neoplasm. Diagnosed as first reported case of primary multiple solitary plasmacytomas affecting bilateral temporoparietal bones in a pediatric patient, the child was treated with external beam radiation therapy to scalp lesions showing excellent results and on further follow-up showed no evidence of progressive disease.

Key words: Multiple solitary plasmacytoma, pediatrics, temporoparietal bones

INTRODUCTION

Plasma cell neoplasms represent a spectrum of diseases characterized by clonal proliferation and accumulation of immunoglobulin-producing terminally differentiated B-cells. In 2003, the International Myeloma Working Group published criteria for the classification of monoclonal gammopathies, multiple myeloma and related disorders recognizing solitary plasmacytoma (SP) of bone, extra-medullary plasmacytoma and multiple SPs (\pm recurrent) as distinct entities.^[1] The majority of plasma cell neoplasms are multiple myeloma, with SP accounting for 6% of cases. The median age at diagnosis of SP is 55-65 years, on average about 10 years younger than patients with multiple myeloma.^[2-4] Males are affected predominately (male:female ratio 2:1).^[2] Multiple SP is called

when more than one localized area of bone destruction or extra-medullary tumor of clonal plasma cells present without any serum M protein elevation or systemic features or organ dysfunction.^[5] Plasmacytomas in the pediatric age group are rare. Here, we are going to present the first reported case of multiple SPs of bilateral temporoparietal bones of a child and response after treatment.

CASE REPORT

A 9-year-old beta thalassemia female child was referred to tertiary cancer center for suspected scalp tumor. Relevant clinical findings included moderate pallor, icterus, typical hemolytic facies, growth delay, tachycardia with hemic murmur and distended abdomen. Spleen was enlarged, non-tender, smooth surface, sharp border, 18 cm along splenic axis with hepatomegaly 10 cm along the mid-clavicular line, smooth surface, tense, sharp margin. Examination of scalp revealed two masses, a mass over left parietal bone, 14.5 cm \times 12.5 cm \times 4.6 cm, soft, non-tender, free from bone, smooth surface, rounded margin, non-fluctuant, and second mass overlying right temporal bone 9 cm \times 8 cm \times 2.7 cm, soft, non-tender, free from bone, smooth surface, rounded margin, non-fluctuant. Contrast enhanced computed tomogram of the brain showed an

Access this article online

Quick Response Code:



Website:

www.cci-journal.org

DOI:

10.4103/2278-0513.119255

Address for correspondence: Dr. Krishnangshu Bhanja Choudhury, Department of Radiotherapy, RG Kar Medical College and Hospital, Kolkata, India. Residence: C-11/4, Green Tower, Golf Green, Kolkata - 700 095, West Bengal, India. E-mail: krishnangshuchoudhury@gmail.com

irregular bony erosion at left parietal area with underlying scalp hematoma, suggestive of tubercular osteitis or neuroblastoma. Mantoux test was non-reactive. Skiagram of the chest showed mild cardiomegaly. Ultrasonography of the abdomen showed huge hepatosplenomegaly. Fine needle aspiration cytology (FNAC) of the left parietal mass showed the presence of abundant plasmacytoid cells some of which are binucleated along with free erythroblasts. Cytological features were suggestive of plasmacytoma [Figure 1]. Repeat FNAC from the swelling over the left side of the head showed predominantly disposed population of cells. Sheets of plasma cells mostly mature are seen admixed with few erythroblasts and myeloblasts, suggestive of plasmacytoma. No M band was detected on serum protein electrophoresis. Urine was negative for Bence-Jones protein. The child was reactive for human immunodeficiency virus (HIV) 1 and 2 but not on anti-retroviral therapy and non-reactive for hepatitis B virus surface antigen and anti hepatitis C virus antibody. Bone marrow aspiration showed typical features of chronic haemolytic anaemia with increased marrow iron store, increased erythroid myeloid ratio with plasma cell less than 5%. The child was diagnosed with multiple solitary extra-medullary plasmacytomas. The child was immobilized by thermoplastic cast and base plate and neck rest B. Planning digital imaging and communications in medicine computed tomography was and data transferred to ASHA treatment planning system. Contouring of visible lesion carried out following the international commission on radiation units and measurements recommendations and 2.5 cms margin in all direction given to mark the clinical target volume. Oblique beams were arranged both side of the skull to cover CTV [Figure 2]. Both masses received radiation dose of 50 Gray in conventional fractionation over 5 weeks. The patient was on follow-up at 3 monthly interval with complete hemogram, serum creatinine, albumin, lactate dehydrogenase, β 2micoglobulin, 24 hr urine for total protein and serum protein electrophoresis. 1 year into the follow-up patient had no local disease recurrence or did not develop multiple myeloma.

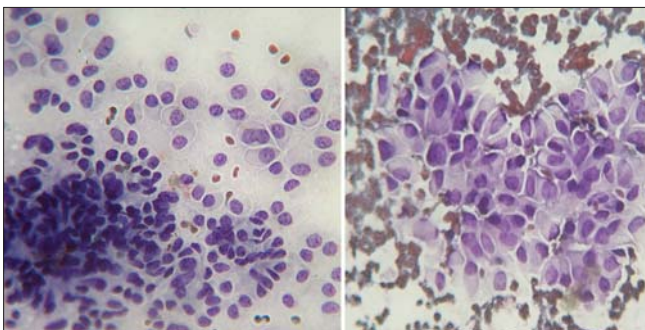


Figure 1: Microphotographs showing cytomorphology of atypical plasma cells, often binucleated, in plasmacytoma, Pap smear $\times 400$

DISCUSSION

Plasmacytoma may be primary or secondary to disseminated multiple myeloma, osseous (medullary) or non-osseous (extra-medullary) in origin and solitary or multiple. While multiple myeloma is extremely rare in children accounting for less than 1% of all patients with myeloma, the incidence of solitary plasmacytomas of bones or extra-medullary plasmacytomas in the pediatric age group are even more rarely reported. Isolated cases affecting base of skull, lymph nodes, and head and neck region have been reported. Involvement of base of skull, arising from the dura matter and involving the dura of left cerebral convexity, tentorium cerebelli and dura of posterior fossa in a 7-year-old child have been reported in early nineties in published case report who eventually progressed to multiple myeloma within 2 years.^[5] In a published case report in 2012, an 11-year-old girl, reportedly suffering from well-differentiated immunoglobulin A/kappa plasmacytoma at the base of the skull at 9 years of age and with negative bone marrow biopsy, ultimately progressed to frank multiple myeloma within 2 years.^[6] The bone marrow biopsy showed a well-differentiated (Marschalko-type) multiple myeloma that was positive for cell surface cluster of differentiation biomarker 138 and immunoglobulin A, with kappa light chain restriction. Interestingly, the Epstein-Barr virus (EBV) was detected by *in-situ* hybridization for EBV-encoded ribonucleic acid in the majority of the neoplastic cells from the biopsy specimens. The patient had responded favorably to treatment with dexamethasone, thalidomide, and zoledronic acid and was scheduled for bone marrow transplantation as per the case report.

Extracting data from plasmacytoma in the adult population, skull involving the cranial vault, skull base and the orbit and the central nervous system are rarely involved by plasma cell tumors without evidence of a plasma cell dyscrasia at

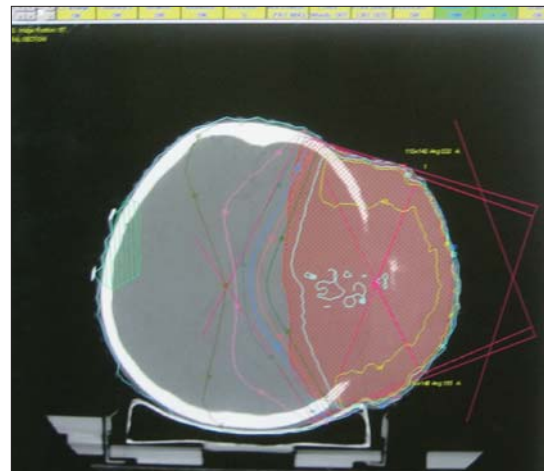


Figure 2: Radiation portals during external beam radiotherapy planning

another site.^[7] The treatments in children have largely been based on the studies in the adult population. The optimal therapy for true SP is curative dose 40-50 Gray radiotherapy. With this radiation dose, local tumor recurrence rate is less than 10%; 30% of patients with solitary bone lesions versus more than 70% of patients with solitary extra-medullary plasmacytomas achieve long disease free survival.

CONCLUSION

Here, we report a rare case of highly radiosensitive, multiple SPs arising from skull bones in HIV seropositive thalassemia child which had shown excellent local control with radiation therapy.

REFERENCES

1. International Myeloma Working Group. Criteria for the classification of monoclonal gammopathies, multiple myeloma and related disorders: A report of the International Myeloma Working Group. *Br J Haematol* 2003;121:749-57.
2. Ozsahin M, Tsang RW, Poortmans P, Belkacémi Y, Bolla M, Dinçbas FO, *et al.* Outcomes and patterns of failure in solitary plasmacytoma: A multicenter rare cancer network study of 258 patients. *Int J Radiat Oncol Biol Phys* 2006;64:210-7.
3. Strojjan P, Soba E, Lamovec J, Munda A. Extramedullary plasmacytoma: Clinical and histopathologic study. *Int J Radiat Oncol Biol Phys* 2002;53:692-701.
4. Tsang RW, Gospodarowicz MK, Pintilie M, Bezjak A, Wells W, Hodgson DC, *et al.* Solitary plasmacytoma treated with radiotherapy: Impact of tumor size on outcome. *Int J Radiat Oncol Biol Phys* 2001;50:113-20.
5. Dincer C, Baskaya M, Yucer N. Solitary intracranial plasmacytoma in a child. Report of a 7-year-old girl. *Turkish Neurosurg* 1994;4:47-50.
6. García-Álvarez KG, Garibaldi-Covarrubias R, Flores-Márquez MR, Ortiz-Hidalgo C. Plasma cell myeloma associated with Epstein-Barr virus infection in an 11-year-old girl. *Pediatr Dev Pathol* 2012;15:339-42.
7. Dimopoulos MA, Moulopoulos A, Delasalle K, Alexanian R. Solitary plasmacytoma of bone and asymptomatic multiple myeloma. *Hematol Oncol Clin North Am* 1992;6:359-69.

Cite this article as: Mallick SK, Chatterjee D, Majumder D, Choudhury KB. A case of plasmacytoma of scalp in pediatric thalassemia human immunodeficiency virus positive child. *Clin Cancer Investig J* 2013;2: 260-2.

Source of Support: Nil, **Conflict of Interest:** None declared.