CORRESPONDENCE

Horner's Syndrome in a Case of Granulocytic Sarcoma

Aditya Jandial¹ · Ram V. Nampoothiri¹ · Pankaj Malhotra¹ · Paramjeet Singh² ·
Man Upadesh Singh Sachdeva³ · Subhash Varma¹

Received: 13 October 2016 / Accepted: 24 October 2016 / Published online: 1 November 2016
© Indian Society of Haematology & Transfusion Medicine 2016

Horner's syndrome is a unique neurological syndrome caused by interruption of the sympathetic supply to one half of the face including the ipsilateral eye. Irrespective of the location of the interruption of sympathetic pathway, Horner's syndrome typically presents with a combination of ipsilateral miosis, partial ptosis and facial anhidrosis. It is primarily a clinical diagnosis which may be confirmed by pharmacological testing. Etiology can be broadly categorised depending upon the neuron involved (first, second or third order). A systematic approach to diagnostic evaluation of Horner's syndrome is essential as the causes are heterogeneous and may range from benign to life threatening. We present a 50-years-old lady, with no previous comorbidities, who consulted a local physician for complaints of reduced sweating of right side of the face, drooping of right upper eyelid and right upper non anginal chest pain radiating to right upper limb. She also had low grade fever and easy fatigability for 15 days. Routine investigations revealed anemia (hemoglobin—8.5 g/dL), thrombocytopenia (23,000/mm³) and leucocytosis (total leucocyte count—22,900/mm³) with 29% blasts in peripheral smear. She was referred to our institute as a suspected case of acute leukemia. Physical

examination at admission at our center revealed pallor. She was afebrile and her blood pressure was 120/84 mm of mercury and pulse rate of 102 per minute. She had right sided partial upper eyelid ptosis, miosis and apparent enophthalmos (Fig. 1). Rest of neurological examination was normal. The presence of ipsilateral hemifacial anhidrosis suggested extra-axial preganglionic right sided Horner's syndrome. Rest of the systemic examination was unremarkable.

Bone marrow examination revealed hypercellular marrow with 49% blasts which contained pale granular basophilic cytoplasm and Auer rods. Staining for myeloperoxidase was positive and overall features were consistent with acute myeloid leukemia. Flow cytometry was suggestive of acute myelomonocytic leukemia with positivity for CD13, CD33, CD117, CD34, CD45, HLADR, CD38, CD19 and CD4. Molecular genetic analysis revealed presence of translocation (8;21) (AML-ETO1). Contrast enhanced MRI of brain and cervico-dorsal spine showed T1 and T2 hypointense, contrast enhancing right paravertebral mass extending from C7 to D4 level into the epidural space (Fig. 2a, b). Brain and spinal cord were normal on MRI. CT guided fine needle aspiration from the mass revealed features consistent with granulocytic sarcoma (myeloperoxidase positive).

Patient chose to receive treatment in the form of hypomethylating agents. She was administered first dose of Inj. Azacytidine 100 mg/m² (175 mg) in hospital. She tolerated it well and was discharged. Commonly described malignancies associated with Horner's syndrome are lung carcinoma, neuroblastoma, thyroid carcinoma, Burkitt's lymphoma and Hodgkin's disease [1–3]. Horner's syndrome in hematology practice has only been rarely described. Non-Hodgkin's lymphoma is the most common hematological malignancy causing

✉ Pankaj Malhotra
hematpgi@gmail.com

¹ Clinical Hematology Division, Department of Internal Medicine, Postgraduate Institute of Medical Education and Research, Chandigarh 160012, India

² Department of Radio Diagnosis and Imaging, Postgraduate Institute of Medical Education and Research, Chandigarh 160012, India

³ Department of Hematology, Postgraduate Institute of Medical Education and Research, Chandigarh 160012, India

Fig. 1 Clinical photograph showing incomplete ptosis in the *right eye*, right pupil miosis (in comparison to right pupil) and enophthalmos of the *right eye*

Horner's syndrome as a result of involvement of cervical lymph nodes, lung apex, thyroid gland and intramedullary spinal cord [1, 4, 5]. We did the literature search through PUBMED using keyword “acute leukemia”, “acute myeloid leukemia” AND “Horner's syndrome” to find the frequency of association. Acute myeloid leukemia, has never been reported to be associated with Horner's syndrome at diagnosis [6]. Horner's syndrome with peripheral neuropathy has been reported in a case of acute promyelocytic leukemia who received high dose cytarabine [7]. In another case of acute myeloid leukemia, Horner's syndrome developed as a part of Pancoast

syndrome due to pulmonary mucormycosis in a setting of febrile neutropenia post chemotherapy [8]. Two cases of acute lymphoid leukemia associated with Horner's syndrome at diagnosis have been described in literature [9, 10]. The presence of myelomonocytic variant (AML M4) of AML and the presence of translocation (8;21) in the index case is consistent with findings of earlier studies. Acute myelomonocytic leukemia (AML M4) and acute monocytic leukemia (M5) are the FAB subtypes most frequently associated with extramedullary manifestations [11] while translocation (8;21) is found to be a recurrent aberration in myeloid sarcomas esp orbital granulocytic sarcomas in children [12]. The overall incidence of extramedullary leukemia in AML has been estimated about 9%, higher incidence described in younger age groups.

In conclusion, haematological malignancies are rare causes of Horner's syndrome and granulocytic sarcomas in AML presenting as Horner's is even rarer. A comprehensive history and physical examination are indispensable components in the approach to a case of Horner's syndrome for localization of lesion and for clues towards etiology. Myeloid sarcoma should be considered in the list of differential diagnoses for a paravertebral soft tissue mass. Fine needle aspiration of the mass with immune-cytochemistry and magnetic resonance imaging are essential investigations for confirming the etiology in a case of Horner's syndrome in such a scenario.

Fig. 2 **a** T1—weighted magnetic resonance imaging of the cervical spine hypointense right paravertebral mass with extension into the epidural space. **b** T2—weighted magnetic resonance imaging of the cervico-dorsal spine sagittal section showing the same mass

Compliance with Ethical Standards

Conflict of interest The authors do not have any conflicts of interests to disclose.

Informed Consent The authors state that written consent was obtained from the patient for publication of any material relating to her including photographs.

Ethical Standards The authors declare that the article follows the required ethical standards as per Helsinki's declaration.

References

1. Nissenbaum M, Kaban LB, Troulis MJ (2007) Toothache, paresthesia, and Horner syndrome: an unusual presentation of disseminated Burkitt's lymphoma. *J Oral Maxillofac Surg* 65(7):1395–1401
2. Lee JH, Lee HK, Lee DH, Choi CG, Kim SJ, Suh DC (2007) Neuroimaging strategies for three types of Horner syndrome with emphasis on anatomic location. *AJR Am J Roentgenol* 188(1):W74–W81
3. Walton KA, Buono LM (2003) Horner syndrome. *Curr Opin Ophthalmol* 14(6):357–363
4. Ruiz ERLS, Gaiolla RD, Niero-Melo L, Custodio Domingues MA, de Lima Resende LA (2012) Post-ganglionic Horner's syndrome: an unusual presentation of non-Hodgkin lymphoma. *Case Rep Neurol* 4(1):43–46
5. Rao RD, Robins HI (2001) Non-Hodgkin's tumor and Pancoast's syndrome. *Oncol Rep* 8(1):165–166
6. Yamamoto JF, Goodman MT (2008) Patterns of leukemia incidence in the United States by subtype and demographic characteristics, 1997–2002. *Cancer Causes Control* 19(4):379–390
7. Nevill TJ, Benstead TJ, McCormick CW, Hayne OA (1989) Horner's syndrome and demyelinating peripheral neuropathy caused by high-dose cytosine arabinoside. *Am J Hematol* 32(4):314–315
8. Bansal M, Martin SR, Rudnicki SA, Hiatt KM, Mireles-Cabodevila E (2011) A rapidly progressing Pancoast syndrome due to pulmonary mucormycosis: a case report. *J Med Case Rep* 5:388
9. Fountaine TJ, Miller B, Khalifa YM, Andolina JR (2012) Horner syndrome in a newly diagnosed patient with high risk precursor B-cell acute lymphoblastic leukemia. *Pediatr Blood Cancer* 59(2):344
10. Imrie K, Baker MA, Messner HA, Keating A (1992) Prolonged disease-free survival in an adult presenting with Burkitt-type acute lymphoblastic leukaemia and CNS disease. *J R Soc Med* 85(1):47–48
11. Bakst RL, Tallman MS, Douer D, Yahalom J (2011) How I treat extramedullary acute myeloid leukemia. *Blood* 118(14):3785–3793
12. Pileri SA, Ascani S, Cox MC, Campidelli C, Bacci F, Piccioli M et al (2007) Myeloid sarcoma: clinico-pathologic, phenotypic and cytogenetic analysis of 92 adult patients. *Leukemia* 21(2):340–350