Open Access Journal

Indian Journal of Medical Research and Pharmaceutical Sciences

September 2018;5(9) ISSN: ISSN: 2349-5340

DOI: 10.5281/zenodo.1437314 Impact Factor: 4.054

NEUROBLASTOMA PRESENTING AS MULTIPLE SCALP SWELLING: A CASE Report

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Keywords:

Proptosis, scalp swelling, ecchymosis.

Abstract

Neuroblastoma is a tumor derived from primitive cells of the sympathetic nervous system and is the most common solid tumor of childhood. The cause of neuroblastoma is unknown in most causes. Neuroblastoma usually occurs sporadically, but familial incidenes have been reported. It is primarily a tumor of abdominal origin from which it metastasizes to liver, lymph nodes, cranium, orbit and to other sites. The prognosis of this disease worsens as the age advances. We report a case of neuroblastoma in a 4 year old child who had B/L proptosis and irregular scalp swelling for 2-3 months with sub ocular petechiae and ecchymosis.

Introduction

Neuroblastoma is the 3rd most common neurogenic, extracranial, solid tumor of infancy and childhood emerging anywhere along the peripheral nervous system [1]. It was first described by Dr. Rudolf Virchow [2]. The prevalance of neuroblastoma is approximately 1/7000 live births [3]. It mostly originates from adrenal gland, nerve tissues of neck, chest, abdomen or pelvis [4].

Case Report

A 4yr old female patient presented with chief complaints of irregular fever for 2-3 weeks. Irregular scalp swelling for 2-3 months and swelling of eyes for 1 month. History revealed child had received 1 blood transfusion 2 months back and have consulted local doctors and recieved some medications for fever. The parents medical history is insignificant. She is a second order child of healthy non consanguinous parents.

General examination revealed weak, thin built girl child with bilateral proptosis with some pallor. There is irregular boggy swelling of the scalp which is fluctuant, non-tender. There is hepatosplenomegaly,liver enlarged 4cm below right costal margin ; liver span being 13 cm and spleen is palpable 3cm below left costal margin. No lymphadenopathy present.

The child was subjected to hematological and radiological investigations which revealed normocytic normochromic anemia with anisopoikilocytosis and isoechoic SOL adjoining upper polar region of Rt kidney in ultrasonography.

Initially based upon clinical presentations the child was diagnosed as case of Langerhan cell histiocytosis but later suspected to be an abdominal malignancy and thus was subjected to bone marrow examination.

The bone marrow aspiration cytology was done in our medical college pathology department which revealed metastatic round cell tumor neuroblastoma. Due to the lack of facility of immunohistochemistry, malignant round cell tumor was not subjected to immunohistochemical examination and was planned to be treated in line of neuroblastoma. After counselling the prognosis our patient was referred to Hematooncology department,AIIMS, Bhubaneswar for further management.

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Discussion

Neuroblastoma has varied clinical presenation and mimicks many other diseases and hence becomes a challenge for diagnosis in a resource limited setting [5].

The clinical presentation reflects the tumor's primary location and extent of metastatic disease, if present [6]. The most common primary for neuroblastoma is abdomen which may metastasize to bone, lymph node, brain, orbit, lungs [7].Clinically neuroblastoma can present with proptosis, periorbital ecchymosis, bone pain, hypertension, watery diarrhoea, fever, anemia, subcutaneous skin nodule. In present case metastasis is rapid involving orbit, abdomen and skull bones [8].

Neuroblastomas have very broad spectrum of clinical behaviour which ranges from spontaneous regression to maturation of a benign ganglioneuroma or aggressive disease with metastasis causing death [9]. As per international classification of childhood neuroblastoma the prognosis is based on the patients age of diagnosis. Children over 5 years usually have a poor prognosis [10]. Based on these tools the prognosis of my Patient will be less than 50%. Treatment. options are surgical resection and chemotherapy for low risk patients. In high risk cases combination of high dose chemotherapy , surgical resection, radiation therapy and stem cell therapy is needed. But eventually despite of the combination therapy the patients have relapses and die of disease [11]. Multiple scalp swellings along with hepatosplenomegaly without mass abdomen always gives a clue towards Langerhan's cell histiocytosis. B/L proptosis can be a feature of Acute Leukemia, but multiple irregular swellings of scalp was not explained by Leukemia. But Getting evidence of small suprarenal tumor (Rt) on USG Abdomen high possibility of Neuroblastoma was thought off. Multiple scalp swellings were due to metastasis from the primary tumor site in the abdomen.

Conclusion

Neuroblastoma in older than 5yrs carries a poor prognosis. A multi disciplinary approach is needed to diagnose and treat the patients. Multiple scalp sewlling though commonly attributed by LCH can also be a manifestation of cranial metastasis of Neuroblastoma .Varied clinical presentation can be present hence all branches should be aware of this disease and have a high index of suspicion for early diagnosis management of these patients.

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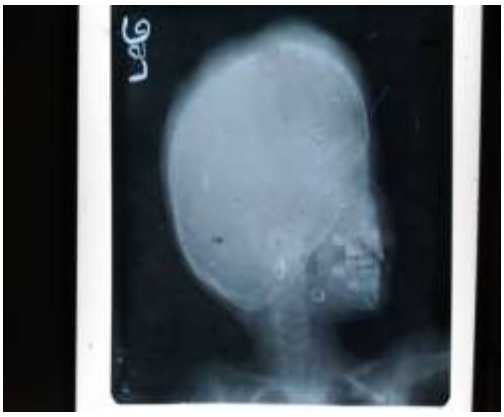

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