Case Report

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Unraveling the Case of Neuroblastoma

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Abstract

Neuroblastoma is the common extracranial solid tumor in children.Very often they are described as "enigmatic tumor" for its broad spectrum of clinical presentation and biological features. Prognosis varying from clinically incidence benign to unresectable or metastic disease with very poor outcomes. They may present with proptosis, periorbital ecchymosis, abdominal distension, bone pain, pancytopenia, fever, anemia, hypertension, paralysis, watery diarrhea, subcutaneous skin nodule and also with paraneoplastic syndromes. Very rarely they may present as a case of Fever of Unknown origin

Keywords: Neuroblastoma; FUO; Metastasis.

INTRODUCTION

Fever is the most common reason for a child to seek medical care. "Fever of unknown origin (FUO)" was first described in 1961 by Petersdorf and Beeson.[1] FUO was defined as well documented fever of at least 3 weeks duration without an apparent source after 1 week of investigation or three outpatient visit. Although there is no standard definition of pediatric FUO, fever lasting anywhere from 10 days to 3 weeks is generally accepted as the working definition of FUO in children.[2,3]

Infectious diseases are the commonest cause of FUO. Non infectious causes like, malignancy, collagen vascular disorders and auto immune disorders must be searched for as causes of FUO. This is a case of malignancy a case of malignancy presenting as FUO.

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CASE REPORT

2 year old developmentally normal girl child presented with complaints of peri orbital puffiness for 4 weeks, fever for 10 days and pain & swelling over right knee joint for past 4 days.

Past history of child was unremarkable. She was 1st child of non consanguineous marriage with uneventful antenatal history. She was born at term by LSCS with birth weight of 2.6 kg. She was immunized as per the age.

On examination, child was active, alert, vitals stable with pallor and mild peri orbital puffiness. No lymphadenopathy or rash over the body. Right knee joint examination was within normal limits except for minimal swelling. GIT examination showed palpable liver 3 cm below right costal margin with soft to firm in consistency and spleen was not palpable.

Blood investigation showed microcytic hypochromic anemia with other two cell lineage within normal limits, with elevated ESR and CRP. RFT, LFT, SE, urine routine examinations were within normal limits. USG abdomen and right knee joint was within normal limits. ANA was negative. Urine VMA was also normal. Chest x-ray, Xrays of skull and right knee joint were normal.

Bone marrow aspiration showed malignant round cell neoplasm. Immunohistochemistry of BM sample was positive for synaptophysin and chromogranin, suggestive of neuroblastoma. CT Abdomen done to look for primary and found to have 2x1 cm calcified adrenal mass with multiple para-aortic lymphnodes (Fig. 1).

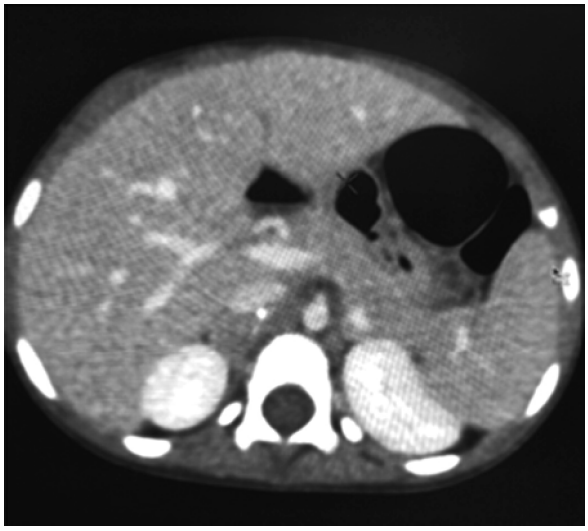

Fig. 1: CT abdomen showing adrenal mass with calcification.

Bone scan showed increased uptake in distal femur and skull suggestive of metastasis (Fig. 2, 3).

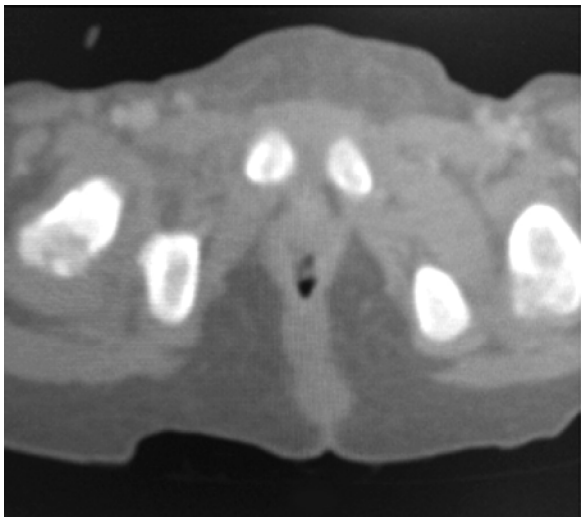

Fig. 2: CT femur showing erosion in bilateral trochanteric areas.

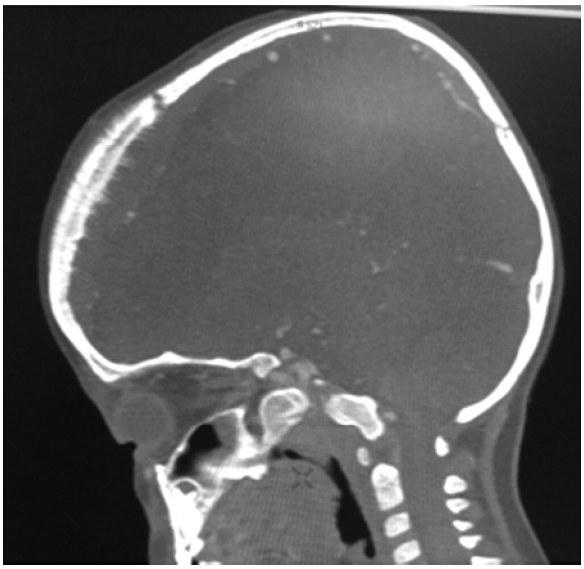

Fig. 3: Skull showing perpendicular periosteal reaction in frontal bone.

Child was referred to higher centre for further management and started with chemotherapy.

DISCUSSION

Neuroblastoma is the common extracranial solid tumor in children. The prevalence is about 1 case per 7,000 live births. It arises from primitive neuroblasts of the embryonic neural crest, and it can occur anywhere within the sympathetic nervous system. The most common primary for neuroblastoma is abdomen which may metastasize to bone, lymph nodes, liver, intracranial, orbital sites, lung, and the central nervous system.[4] Nearly, 50% of the neuroblastomas are diagnosed in children younger than 5 years of age.[5] Neuroblastoma is very often called "enigmatic tumor" for its broad spectrum of clinical presentation, biological features and prognosis varying from benign to unresectable benign to unresectable or metastic disease with very poor outcomes.

Neuroblastoma appear most often in abdomen, it can located anywhere in the body along the peripheral sympathetic nervous system.[6] The presenting signs and symptoms are highly variable. They are related to the site of the primary tumor. Clinically, neuroblastoma presents with proptosis, periorbital ecchymosis, abdominal distension, bone pain, pancytopenia, fever, anemia, hypertension, paralysis, watery diarrhea, and subcutaneous skin nodule.[7]

Majority of patients present with evidence of haematogenous metastases to distant sites such as cortical bone, bone marrow, liver and non regional

lymph nodes.[8] Skeletal metastases occur in up to 60% of cases with a variable radiological appearance. Metastases to the skull are frequent. Skeletal lesions in long bones may present radiographically as osteolytic focus with or without periosteal reaction, lucent horizontal metaphyseal line or vertical linear radiolucent streaks in the metadiaphysis. Very rarely will present with features of paraneoplastics yndromes. (Table 1)

Table 1: Neuroblastoma presenting with features of paraneoplastic syndromes.[9]

Owing to excessive catecholamine (VMA,VHA)	Sweating, flushing, pallor, headache, palpitation
Owing to vasoactive intestinal peptides(VIP)	Dehydration, hypokalaemia and abdominal distention, secretory diarrhoea, failure to thrive (Kerner–Morrison syndrome)
Acute myoclonic encephalopathy (opsoclonus-myoclonus [OM] syndrome)	Opsoclonus (dancing eyes syndrome), myoclonus (irregular jerking of muscles of limbs and trunk)

Most neuroblastomas occur sporadically; around 1-2% are familial and associated with multiple primary tumours, usually occurring at <18 months of age. Familial disease has the same diverse clinical behaviour as somatic neuroblastoma, ranging from aggressive progression to spontaneous regression. Neuroblastoma can be associated with several inborn conditions, like Hirschsprung disease, congenital central hypoventilation syndrome (CCHS or Ondine's curse), neurofibromatosis type I, or Beckwith Wiedemann syndrome.

The most important clinical prognostic variables in patients with neuroblastomas are age at diagnosis, stage of the disease, and the site of the primary tumor.[10] As the age advances, prognosis becomes poor.

Neuroblastoma presenting with isolated fever is rare. USG scan and X-rays of bones may be normal. So higher imaging studies warranted.

REFERENCES

1. Petersdorf RG, Beeson PB. Fever of unexplained origin: report on 100 cases. Medicine (Baltimore), 1961, 40:1-30.
2. J.A. Finkelstein, C.L. Christiansen, R. Platt Fever in pediatric primary care: occurrence, management, and outcomes; Pediatrics, 105 (2000), pp. 260-266
3. A. Chow, J.L. Robinson, Fever of unknown origin in children: a systematicreview; World J Pediatr, 7 (2011), pp. 5-10
4. DuBois SG, Kalika Y, Lukens JN, Brodeur GM, Seeger RC, Atkinson JB, et al. Metastatic sites in stage IV and IVS neuroblastoma correlate with age, tumor biology, and survival. J Pediatr HematolOncol. 1999;21:181–9.
5. Podda MG, Luksch R, Polastri D, Gandola L, Piva L, Collini P, et al. Neuroblastoma in patients over 12 years old: A 20-year experience at the IstitutoNazionaleTumori of Milan. Tumori. 2010;96:684–9
6. PapaioannouG,McHugh K, Neuroblastoma in childhood. Review and radiological findings. Cancer Imaging 2005;5: 116-127
7. Citak C, Karadeniz C, Dalgic B, Oguz A, Poyraz A, Okur V, et al. Intestinal lymphangiectasia as a first manifestation of neuroblastoma. Pediatr Blood Cancer. 2006;46:105–7
8. Chu CM, Rasalkar DD, Hu YJ, Cheng FW, Li CK, Chu WC. Clinical presentations and imaging findings of neuroblastoma beyond abdominal mass and a review of imaging algorithm. Br J Radiol. 2011;84(997):81-91. doi:10.1259/bjr/31861984
9. Lonergan GJ, Schwab CM, Suarez ES, Carlson CL. Neuroblastoma, ganglioneuroblastoma, and ganglioneuroma: radiologic-pathologic correlation. Radiographics 2002;22:911–34
10. Brodeur GM, Castleberry RP: Neuroblastoma. In Principles and Practice of Pediatric Oncology, 2nd ED. Pizzo PA, Poplack DG, EDS. Philadelphia: Lippincott Publisher, 1993, pp 739-67