

Case Report

Physical examination a boon-raccoon eyes in a child with neuroblastoma

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Received: 07 March 2024

Revised: 03 April 2024

Accepted: 08 April 2024

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ABSTRACT

Metastatic neuroblastoma often presents with prolonged fever, cytopenias and significant constitutional symptoms, before the abdominal mass is evident clinically. Clues like subcutaneous nodules and racoon eyes can help a clinician to suspect and evaluate appropriately to arrive at a timely diagnosis. We report a 3-year-old girl who had prolonged fever and constitutional symptoms for more than 2 weeks. On evaluation in outpatient visits, child was found to have cytopenia, hence was referred to our center for further management. Clinical examination revealed raccoon eyes with scalp nodules, hepatomegaly and abdominal mass predominantly over left lumbar area crossing over midline. On further investigations including radiological imaging and bone marrow biopsy child was diagnosed with metastatic neuroblastoma. Metastatic neuroblastoma often presents with prolonged fever, cytopenias and significant constitutional symptoms, before the abdominal mass is evident clinically. Clues like subcutaneous nodules and racoon eyes can help a clinician to suspect and evaluate appropriately to arrive at a timely diagnosis.

Keywords: Raccoon eyes, Scalp nodules, Neuroblastoma

INTRODUCTION

Neuroblastoma is a common childhood cancer especially those under 5-years old; however they can have varied presentations that can make diagnosis a difficult task.¹ This can rapidly metastasize and widely spread, resulting in a variety of clinical manifestations, such as proptosis, periorbital ecchymosis, abdominal distension, anemia, pancytopenia, bone pain, paralysis and numerous Para neoplastic syndromes such as cerebellar ataxia and opsoclonus myoclonus syndrome.² We report a young child who had raccoon eyes and scalp nodules subsequently diagnosed as neuroblastoma.

CASE REPORT

A 2-year-old girl child presented with history of fever for 2 months with irritability, reduced activity and refusal to walk for 2 weeks. Physical examination revealed pallor, periorbital ecchymosis and multiple scalp nodules (Figure 1). Blood pressure was normal. Abdomen examination revealed hepatomegaly and a diffuse hard non-tender mass palpable of size 10×8 cm predominantly in left lumbar region crossing midline. Hemogram revealed hemoglobin of 4.3 gram %, total leucocyte counts of 5370 cells/mm³ and platelet count of 164×10³cells/mm³. Peripheral smear revealed normocytic normochromic anemia. Erythrocyte sedimentation rate (ESR), lactate

dehydrogenase (LDH) and serum ferritin were elevated as 140 millimeter/hour, 1616 unit/litre and 2858 nanogram/ml respectively. Tumor lysis workup was negative. Ultrasound abdomen revealed hepatomegaly, minimal ascites and an ill-defined heteroechoic mass lesion with calcification in the left adrenal region crossing the midline encasing abdominal aorta, coeliac and superior mesenteric vessels. Histopathological examination (HPE) of scalp nodule showed tumor cells with scant cytoplasm, small round nucleus with salt and pepper chromatin arranged in sheets, with occasional apoptotic figures and few mitoses. Immunohistochemistry (IHC) was positive for synaptophysin and chromogranin, and negative for CD45 and NKX2.2, suggestive of metastatic deposit of neuroblastoma (Figure 2 A-C). Positron Emission Tomography (PET) scan done revealed a large heterogeneously enhancing mass lesion with variable FDG uptake, lobulated margins, central necrosis and calcifications is seen in the retro peritoneum measuring 11.5×8.9×10.8 cm; SUV max-4.91. There were also ill-defined lytic lesions noted in skull with significant enhancing soft tissue metastatic lesions in bilateral subdural regions. It showed multiple FDG avid mixed lytic sclerotic lesions in cervical, lumbar vertebrae, pelvic bones, sternum, ribs, scapulae, bilateral humerus and angle of mandible on right side. Few hypodense FDG avid lesions noticed in both lobes of liver suggestive of metastasis. Risk stratification of the patient was done and patient was categorized as high-risk neuroblastoma and was started on chemotherapy as per SIOPEN protocol. Child showed clinical and radiological improvement initially with chemotherapy but later noted progression and succumbed to disease.



Figure 1: Raccoon eyes and scalp nodules in the index child.

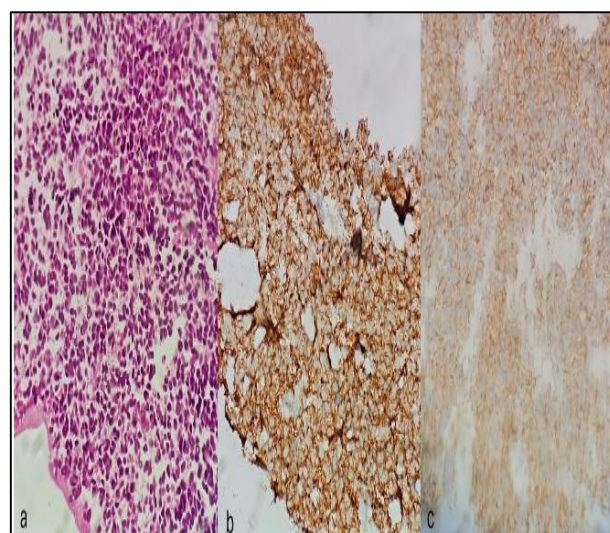


Figure 2 (A-C): Histopathological images showing sheets of infiltrating small blue round cell tumour suggesting a poor differentiated neuroblastoma; immunohistochemistry showing positivity for synaptophysin and chromogranin.

DISCUSSION

Neuroblastoma is a pediatric neuroblastic tumor arising in the sympathetic nervous system from neural-crest cells. Orbital neuroblastoma metastasis is relatively rare and can be seen in up to 20% of children with neuroblastoma, which is associated with poor prognosis. Since the symptoms and signs of orbital neuroblastoma are not specific, its diagnosis remains challenging. Periorbital ecchymosis (raccoon eyes) in a case of neuroblastoma is perhaps related to obstruction of the palpebral vessels (branches of the ophthalmic and facial vessels) by metastatic tissue in and around the orbits. Child abuse, peri-orbital cellulitis, primary systemic amyloidosis, and blood dyscrasias may be considered in the differential diagnosis for raccoon eyes apart from head trauma.³ Clinical manifestations, such as peri-orbital ecchymosis and proptosis, in combination with radiological analysis and histological findings, are important for the diagnosis of orbital neuroblastoma metastasis.⁴ Neuroblastoma, the most common extra cerebral solid tumor of childhood, accounts for the most common tumor metastasizing to the skull in pediatric population. Thus, if a “hair-on-end” skull does occur in children younger than age 5 years, a calvarial metastasis, particularly from neuroblastoma, should be considered as a differential diagnosis.⁵ Skull metastasis has been found in up to 25% of patients with neuroblastoma. These calvarial lesions frequently enlarge to produce epidural deposits. Skull metastases are often very subtle. These calvarial metastases are frequently associated with scalp lesions. Metastatic involvement of the skull in neuroblastoma has various possible radiographic findings: thickened bone, the “hair-on-end” periosteal reaction, lytic defects, and sutural separation. The sutural separation caused by direct involvement of

neuroblastoma differs from that found with generalized increased intracranial pressure. In neuroblastoma, the separation of sutures is not uniform, and the margins of the sutures are partially indistinct.⁶ In a retrospective cohort study on pediatric scalp swellings conducted showed that malignant tumors of the scalp in children were less common, with a total of 5 cases (5/228, 2.19%) in the cohort. Malignancies can often progress rapidly, with imaging characteristics evidencing a poorly defined lytic lesion with an aggressive appearance, extension into bordering soft tissue, and intense heterogeneous enhancement. In addition, for malignant tumors with scalp tumors as the first manifestation, a systemic examination should be performed to determine whether lesions are present elsewhere.⁷

CONCLUSION

A meticulous physical examination is important for early diagnosis and prompt referral of neuroblastoma. Physical examination findings such as raccoon eyes with or without scalp nodules can be a diagnostic clinch in case of neuroblastoma. This soft skill has to be nurtured in the early years of any clinician, which can significantly impact patient outcomes. The above-mentioned case report is a classic example of examination cues leading to provisional diagnosis.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Bhurat R, Siri M, Kandasamy K, Jayaraman D, Scott JX. Physical examination a boon-raccoon eyes in a child with neuroblastoma. Int J Contemp Pediatr 2024;11:593-5.