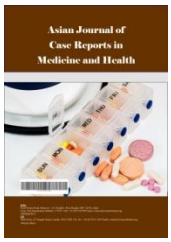

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Neuroblastoma in Early Childhood: A Case Report and Literature Review

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Authors' contributions

This work was carried out in collaboration among all authors. Author HSJ designed the study, wrote the first draft of the manuscript. Authors HAS and MAK managed the literature searches. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Introduction: Neuroblastoma is an extremely rare pediatric neoplasm whose prognosis becomes poor as the age advances. It can be sporadic or nonfamilial in origin. It is primarily a tumor of abdominal origin from where it metastasis to lymph nodes, liver, intracranial and orbital sites, and central nervous system The purpose of this paper is to report a case in which scalp nodules and inguinal lymph node was the initial presenting sign of disseminated neuroblastoma in a 2 year-old child.

Case Presentation: We report a rare case of metastasis of neuroblastoma in a 2-year-old child presenting complain of fever for several weeks, anorexia, loss of weight , arthralgia and multiple scalp nodules and inguinal lymph node enlargement . Biopsy was receive from the scalp and inguinal lymph nodes showed undifferentiated tumor tissue where medium sized atypical blastoid tumor cells were located in clusters and sheets in an eosinophilic sometimes fibrillary background showing blood vessels. The sinuses were also infiltrated by these tumor cells.
Immunohistochemically, Pan Leucocyte antigen was negative as also myogenin and CD99.

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However, CD 56 and synaptophysin are strongly positive and the neuroblastoma marker was weakly positive in the neuropil-like structures and the cytoplasm of some tumor cells.
Conclusion: The addition of our case to the literature offers new clinicopathological data useful for better defining the diagnosis. The differential diagnosis of neuroblastoma in a child presenting with multiple scalp nodules and inguinal lymph node enlargement should be consider.

Keywords: Neuroblastoma; infant; scalp; lymph nodes; metastasi.

1. INTRODUCTION

Neuroblastoma is the third most common neurogenic, extracranial solid tumor of infancy and childhood emerging anywhere along the peripheral sympathetic nervous system [1].

Neuroblastoma accounts for more than 7% of malignancies in patients younger than 15 years and around 15% of all paediatric oncology deaths [2]. It was first described by Dr. Rudolf Virchow as a “glioma” in the abdominal cavity [3]. In 1910 Dr. Homer-Wright presented it as primitive neural cells tumor within the bone marrow [4]. The prevalence of neuroblastoma is approximately 1/7000 live births [5].

Neuroblastoma may be sporadic or nonfamilial in origin6] Exact etiology of neuroblastoma is not well understood, but the recent studies have improved the understanding of genetic susceptibility to neuroblastoma [7]. It originates mostly from the adrenal gland, nerve tissues of the neck, chest, abdomen, or pelvis. [8,9] While 90% of cases are diagnosed before the age of 5, 30% of those are within the first year [10].

The majority of children presenting with neuroblastoma have ‘high risk disease’ with distant metastases at primary diagnosis associated with considerable (more than 50%) mortality [11].

The present case reports neuroblastoma metastasis in scalp nodules and lymph nodes of a 2 year-old boy at the time of diagnosis.

2. CASE REPORT

2-year-old child presenting complained of fever for several weeks, anorexia, loss of weight and arthlagia. There was no history of trauma, convulsions, vomiting, limb weakness, abnormal bleeding or bladder / bowel disturbances.

General examination revealed weak, thin built boy with multiple scalp nodules and inguinal lymph node enlargement, No abnormalities of hand and feet were observed.

The complete blood profile was within normal limits. Chest x-ray was normal. Mantoux test was negative. Urinalysis was within normal limits. There were painless enlarged inguinal lymph nodes.

Skull x-ray revealed bony destruction with irregular margins.

Biopsy was performed from the scalp nodule and inguinal lymph nodes from 1.5 to 2 cm.

Histological lymph node sections showed undifferentiated tumor tissue where medium sized atypical blastoid tumor cells are located in clusters and sheets in a fibrillary background. The sinuses were also infiltrated by these tumor cells. In the scalp nodule tissue there were osteoid rich bone trabeculae that were infiltrated by the same tumor tissue. The PAS stain was negative.

Immunohistochemical; Pan Leucocyte antigen was negative as also myogenin and CD99. However, CD 56 and synaptophysin were strongly positive and the neuroblastoma marker was weakly positive in the neuropil-like structures and the cytoplasm of some tumor cells. The proliferation index at Ki67 is 40%. The lesions was confirmed to be metastasis of neuroblastoma

3. DISCUSSION

Neuroblastoma is the most common malignant solid tumor in the peripheral nervous system in infants and young children, with a high degree of malignancy [12].

The clinical presentation of neuroblastoma reflects the tumor's primary location and the extent of metastatic disease, if present. 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 [13]. Clinically, the neuroblastoma may present with proptosis, periorbital ecchymosis, abdominal distension, bone pain, pancytopenia, fever, anemia, hypertension, paralysis, watery diarrhea, and subcutaneous skin nodule [14].

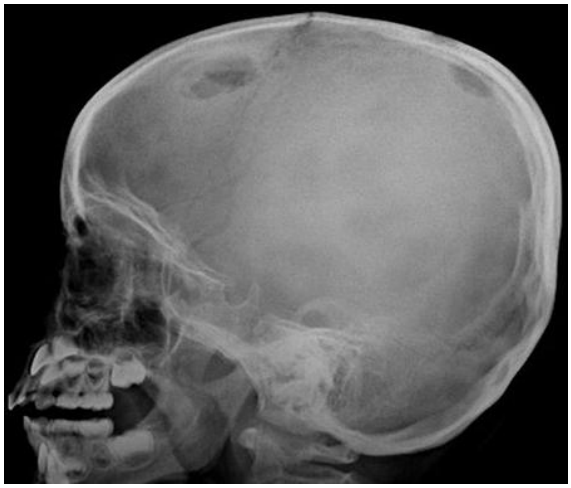

Fig. 1. Skull x-ray revealed bony destruction with irregular margins

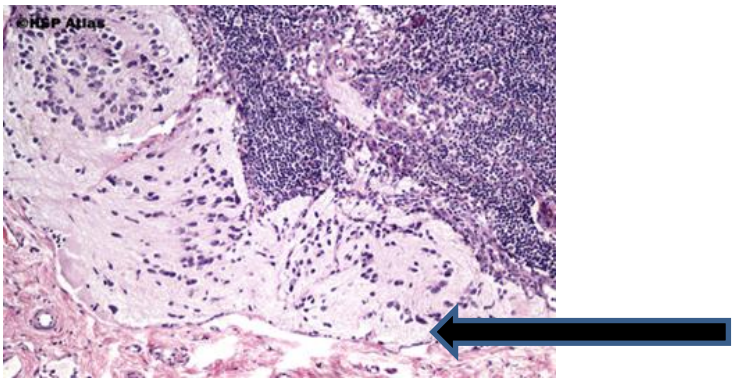

Fig. 2. Histological lymph node sections showed undifferentiated tumor tissue where medium sized atypical blastoid tumor cells are located in clusters and sheets in a fibrillary background. The sinuses are also infiltrated by these tumor cells

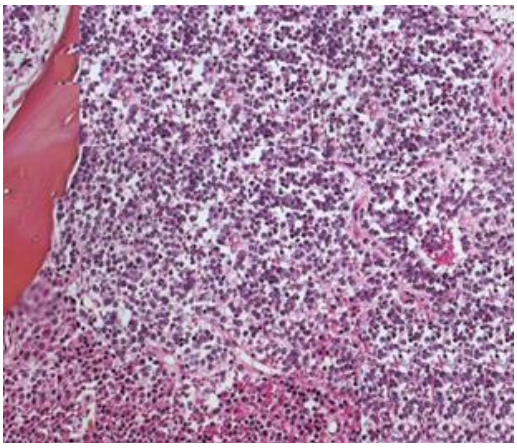

Fig. 3. The scalp nodule tissue there were osteoid rich bone trabeculae that are infiltrated by the same tumor tissue

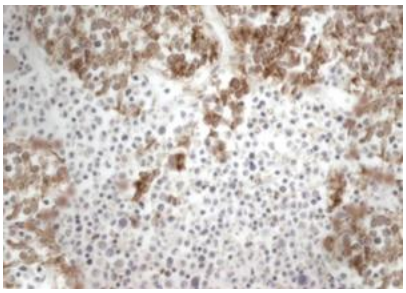

Fig. 4. Synaptophysin Immunohistochemical

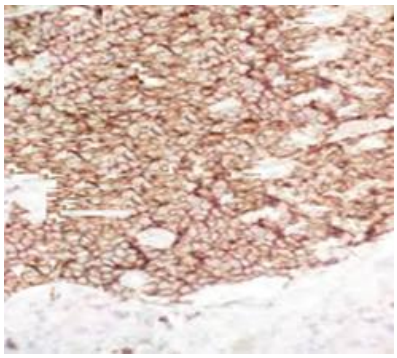

Fig. 5. CD 56 Immunohistochemical Synaptophysin and CD 56 were strongly positive

Sometimes the first symptoms of neuroblastoma are often vague, making diagnosis difficult sometimes they present with paraneoplastic manifestations [15]. In our case the patient at first developed only some vague symptoms, for example fever for several weeks, anorexia, loss of weight, arthralgia .So in any patient of prolonged undiagnosed fever, possibility of a malignancy like neuroblastoma should be kept in mind .Approximately 70% of patients with neuroblastoma present with metastatic disease, [16].

Common migration sites include the lymph nodes, bone marrow, bone, liver, and skin [17].

Skull metastasis has been found in up to 25% [18]. Metastatic involvement of the skull has various possible radiographic findings: Thickened bone, the hair-on-end" periosteal reaction, lytic defects, and suturalseparatio [18]. In our case there was skull metastasis with lytic lesions on skull evidenced by X-ray. But no other bony abnormality was found on imaging of thorax, abdomen, and other long bones.

Regional lymph node metastasis was noted in up to 35% of patients with apparently localised tumors, [19] but in our case no primary tumor was found and there was inguinal lymph node metastasis.

On microscopy, the tumor cells are typically described as small, round, and blue. Rosette patterns (Homer-Wright pseudorosettes) may be seen. Although a number of antigens can be found on neuroblastomas, NSE is the most sensitive. Others are neurofilament protein, S-100, chromogranin, synaptophysin, vasointestinal peptide, [15]. In our case the positivity for CD 56 and synaptophysin confirmed the diagnosis.

4. CONCLUSION

The addition of our case to the literature offers new clinicopathological data useful for better defining the diagnosis.

The differential diagnosis of neuroblastoma in a child presenting with multiple scalp nodules and inguinal lymph node enlargement should be consider.

CONSENT

As per international standard or university standard, patient's consent has been collected and preserved by the authors.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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