ORIGINAL RESEARCH PAPER
Volume - 11 | Issue - 01 | January - 2022 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

NEUROBLASTOMA WITH PARAPARESIS: AN UNCOMMON PRESENTATION

Paediatric Medicine

Dr. Smriti Surana* Post Graduate Resident, Pediatrics, Burdwan Medical College and Hospital. *Corresponding Author

Dr. Arijeet Jana Post Graduate Resident, Radiology, Burdwan Medical College and Hospital.

Dr. Kaustav Nayek Professor, Head of Department, Pediatrics, Burdwan Medical College and Hospital.

ABSTRACT

Neuroblastoma is the third most common childhood tumor, after leukemia and brain tumors, originating from the sympathetic nervous system. We report the case of a 6 month old boy presenting to us with abdominal distention and paraparesis. Diagnosis was made after radiological evaluation and fine needle aspiration cytology from the mass.

KEYWORDS

Neuroblastoma, paraparesis, neoplasms

INTRODUCTION

Neuroblastoma is the most common extracranial tumor of childhood, which usually arises paraspinally in the abdomen or chest from precursors cells of the peripheral (sympathetic) nervous system.[1] It accounts for 7% of all pediatric neoplasms[2]

The early age of presentation and the terminally differentiated nature of cells in the mature autonomic nervous system suggest the possibility of a mutagenic event in the prezygotic germ cells, during embryogenesis or early fetal life, resulting in development of neuroblastoma during the neonatal period, in infancy, or childhood.[3]

The clinical presentation of neuroblastoma depends upon the tumor's primary location and the extent of metastatis, if present.[4]

The most common primary for neuroblastoma is abdomen which may then metastasize to bone, lymph nodes, liver, intracranial, orbital sites, lung, and the central nervous system.[5]

The present case report highlights the extremely rare presentation of paraparesis in a 6 month old child with neuroblastoma.

CASE STUDY

A 6 month old male child, first born of a non-consanguineous marriage, singleton, term pregnancy, presented with inability to move both lower limbs shortly after birth, along with gradual distention of the abdomen for 3 months.

General examination revealed a thin built child with abdominal distention and paraperesis. [Figure 1]. On palpation, liver was palpable 4 cm below midclavicular line, firm in consistency. A mass was palpated in the left iliac fossa, firm, non tender. Tone of bilateral lower limbs was decreased, jerks were diminished in the lower limbs. Other examination findings were within normal limits. No history of trauma, ecchymotic patches, bleeding manifestations.

Preliminary blood investigations revealed normal complete blood counts, liver function tests and kidney function tests. Chest roentgenogram was within normal limits. Xray of bilateral lower limbs and of lumbar spine were also within normal limits.

Ultrasound of the abdomen revealed multiple tiny hyperechoic areas noted in both lobes of liver. The left kidney was seemed to be pushed antero – superiorly by a large hypoechoic heterogenous lobulated retroperitoneal SOL with demonstrable vascularity, suggestive of neuroblastoma. [Figures 2-4]

USG guided FNAC from the mass revealed sheets and clumps of round cells against a background of RBC. Cytological features were suggestive of a round cell tumour. [Figure 5]

MRI LS Spine showed tumor mass which was hypointense in T1 weighted image, and heterogenous hyperintense on T2 weighted image. It also showed heterogenous contrast enhancement. Left and right adrenals were separately visualised confirming extra adrenal origin, likely from paravertebral sympathetic chain. Communication into neural canal could be established. Retroperitoneal lymphadenopathy with hepatic metastasis could be confirmed on contrast imaging. [Figures 6-11]

Due to lack of facility, immunohistochemistry of the round cell tumor could not be done. The child was then referred to higher centre for further investigations and management.

Figure 1 : Photograph showing abdominal mass with floppy lower limbs.

Figure 2: Ultrasound abdomen showing retroperitoneal mass.

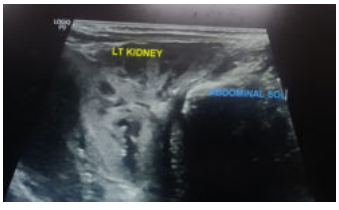

Figure 3 : Ultrasound abdomen showing displacement of left kidney by abdominal mass.

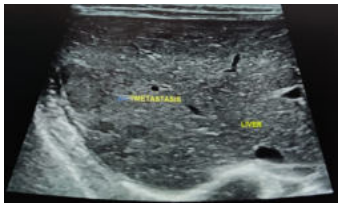

Figure 4 : Ultrasound abdomen showing metastasis to liver

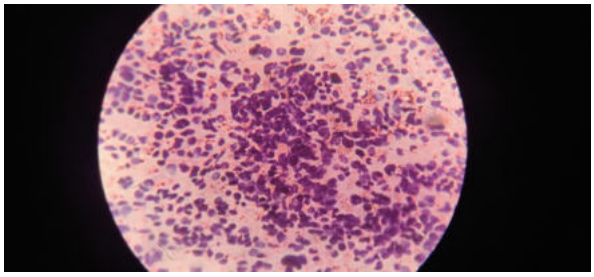

Figure 5: Light microscopic H and E stained picture revealed sheets and clumps of round tumor cells, some of which form rosettes.

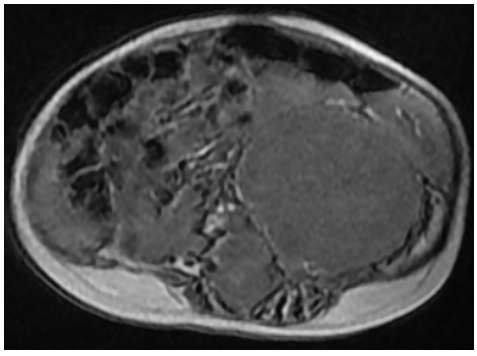

Figure 6 : Axial T1 weighted image showing hypointense lesion

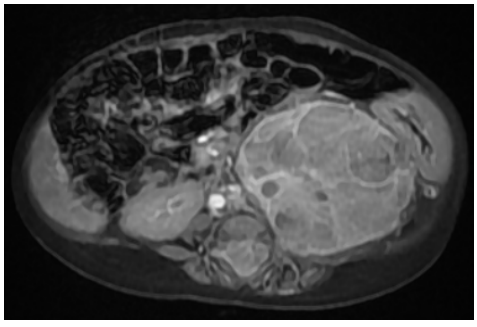

Figure 7: Axial post contrast T1 fat saturated image showing heterogeneous enhancement in left lumbar region, pushing kidney anteriorly

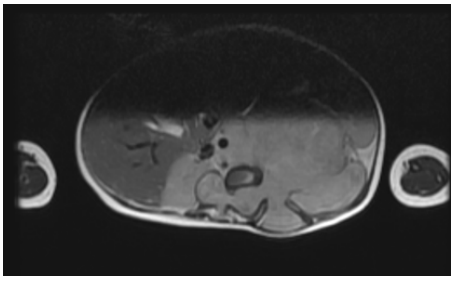

Figure 8 : Axial T2 weighted image showing intraspinal extension of the lesion with anterior displacement of Inferior Vena Cava and Aorta

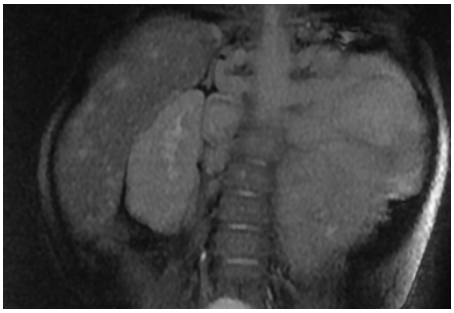

Figure 9 : Coronal T2 weigheted image showing the normal right adrenal gland and iso-hyperintense lesion in the left lumbar region

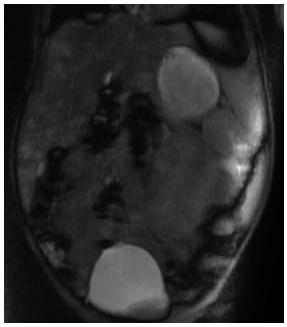

Figure 10 : Coronal T2 weighted image showing normal left adrenal gland.

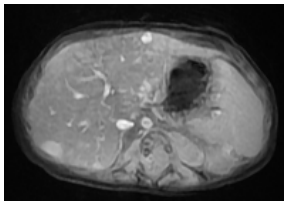

Figure 11 : Axial post contrast T1 fat suppressed image showing hepatic metastasis.

DISCUSSION

Neuroblastomas have a very broad spectrum of clinical progression, ranging from spontaneous regression to maturation of a benign ganglioneuroma, or aggressive disease with metastasis leading to death.[5] Most signs and symptoms of neuroblastoma are attributable to local problems from the primary tumor or are the result of hormone produced by the tumor. Most tumors are hormonally active and produce catecholamines that result in hypertension, diarrhea, dehydration, hypokalemia, skin rashes, and flushing[6]

As per the international classification of neuroblastoma, the prognosis of childhood neuroblastoma is based on the patient's age at diagnosis, deteriorating as the age progresses.[7]

Till date, the management of patients with neuroblastoma remains complex and difficult. In low-risk patient, surgical resection in conjugation with chemotherapy is the first line treatment. For high-risk pediatric patients, aggressive surgical resection high-dose chemotherapy along with stem cell rescue, radiation therapy, and biologic/immunologic therapy have been proposed. Spinal cord involvement is seen in 7 to 15% of children with neuroblastoma[8], and chemotherapy and laminectomy are the modalities of choice for spinal cord decompression[9]. Malignant spinal tumors with spinal compression typically have a poor prognosis, including paralysis, sensory loss, and sphincter dysfunction. Thus, early diagnosis and treatment are essential to minimize morbidity[10-11]. However, incidence of relapse and mortality are very high in these patients.[12]

CONCLUSIONS

Neuroblastoma is a common extracranial solid malignant tumor. Spinal cord compression signs and symptoms are subtle in case of infants. The importance of early recognition and management is important for utmost benefit of patient.

REFERENCES

1. Brodeur GM. Neuroblastoma: biological insights into a clinical enigma. Nature Reviews Cancer. 2003 Mar;3(3):203-16.
2. Becker J. Wilting, J. WNT signaling, the development of the sympathoadrenal–paraganglionic system and neuroblastoma. Cellular and Molecular Life Sciences. 75, 1057–1070 (2018).
3. Chan AR, Johnston DH, Stoneman D. Disseminated neuroblastoma with initial presentation as an intraoral mass: case report. Pediatric Dentistry. 1994 Jul-Aug;16(4):310-3.
4. Kalaskar RR, Kalaskar AR. Neuroblastoma in early childhood: A rare case report and review of literature. Contemporary Clinical Dentistry. 2016;7(3):401.
5. DuBois SG, Kalika Y, Lukens JN, Brodeur GM, Seeger RC, Atkinson JB, Haase GM, Black CT, Perez C, Shimada H, Gerbing R, Stram DO, Matthay KK. Metastatic sites in stage IV and IVS neuroblastoma correlate with age, tumor biology, and survival. Journal of Pediatric Hematology/ Oncology. 1999 May-Jun;21(3):181-9.
6. Wilcox PB, Wright JM, Byrd RL. Metastatic neuroblastoma to the jaws: report of two cases. ASDC Journal of Dentistry for Children. 1984 May-Jun;51(3):211-4.
7. Shimada H, Umehara S, Monobe Y, Hachitanda Y, Nakagawa A, Goto S, Gerbing RB, Stram DO, Lukens JN, Matthay KK. International neuroblastoma pathology classification for prognostic evaluation of patients with peripheral neuroblastic tumors: a report from the Children's Cancer Group. Cancer. 2001 Nov 1;92(9):2451-61.

8. Shohet JM, Nuchtern JG, Lowas SR. Treatment and prognosis of neuroblastoma [Internet]. UpToDate. [cited 2021Dec13]. Available from: https://www.uptodate.com/contents/treatment-and-prognosis-of-neuroblastoma
9. Yahya FS, Al-Shami HA. Posterior mediastinal neuroblastoma masked as flaccid paraparesis in a 3 year child. Neurosciences. 2019;24(4):320–3.
10. De Martino L, Spennato P, Vetrella S, Capasso M, Porfito C, Ruotolo S, et al. Symptomatic malignant spinal cord compression in children: A single-center experience. Italian Journal of Pediatrics. 2019;45(1).
11. Tantawy AAG, Ebeid FSE, Mahmoud MA, Shepl OE. Spinal cord compression in childhood pediatric malignancies. Journal of Pediatric Hematology/Oncology. 2013;35 (3): 232–6.
12. Yalçin B, Kremer LC, Caron HN, van Dalen EC. High-dose chemotherapy and autologous haematopoietic stem cell rescue for children with high-risk neuroblastoma. Cochrane Database Systematic Review. 2013 Aug 22;(8):CD006301.