ISSN: 0975-833X

Available online at http://www.journalcra.com

International Journal of Current Research
Vol. 14, Issue, 09, pp.22229-22231, September, 2022
DOI: https://doi.org/10.24941/ijcr.44004.09.2022

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

RESEARCH ARTICLE

A CASE OF GANGLIONEUROBLASTOMA – DIAGNOSTIC CHALLENGE

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ARTICLE INFO

Article History:
Received 09th June, 2022
Received in revised form
27th July, 2022
Accepted 19th August, 2022
Published online 28th September, 2022

Key words:

Neuroblastoma,
Ganglioneuroblastoma,
Abdominal Mass.

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ABSTRACT

Among pediatric patients, neuroblastoma is the most common tumor of extracranial origin. They are derived from embryonic cell involved in the development of the sympathetic nervous system, whose differentiation has been halted. Most tumors are diagnosed in the first decade of life. Disease development is unpredictable. It ranges from progression of full blown disease or may spontaneously regress. This variation is due to the rate of maturation, location and metastatic potential. In this case study a typical case of Ganglioneuroblastoma and its prognostic significance has been described.

Citation: ***Aarthi Arulvanan, Krishnaswamy, B., Dhanalakshmi, M., Manohar, U. and Shyamala, E.*** **2022.** "A Case of Ganglioneuroblastoma – Diagnostic Challenge*". International Journal of Current Research*, 14, (09), 22229-22231.

INTRODUCTION

An immature neuroblast and schwannian poor stroma constitute Neuroblatoma. Ganglioneuroblastoma is mixture of immature neuroblasts and ganglion cells; schwannian rich stroma. Ganglioneuroma is composed only of ganglion cells; schwannian stroma dominant. It is the 4th most common tumor in childhood. 75-80% are found in children under 5 years of age and rarely occurs in adult[5]. Males are commonly affected than females. The tumor can arise wherever sympathetic nerve is present. The most common location are adrenal gland (35%), para spinal and retroperitoneal ganglia (30-35%), posterior mediastinum, head and neck (1-5%) [1].

CASE REPORT

A 1 year 9 month male child came to hospital with complaint of loss of weight, lethargy, abdominal mass. On examination diffuse swelling in the left para spinal area noted. Ultrasound abdomen was done on 22/9/21 shows 5x6cm sized hyper echoic lesion seen in left para spinal region at the level of dorsal vertebra, communicating with spinal. CECT abdomen was done on 23/9/21 shows 6X7cm left para vertebral mass with dumbbell extension to D12-L3 spinal cord. Bony erosion is seen.MRI abdomen shows Left para vertebral mass 8x4x6cm extending into spinal canal compressing spinal cord. Staged procedures of Laprotomy followed by posterior laminectomy D12-L3 was done to remove the tumor. Intraop findings: Left adrenal mass of 3x3cm along with paravertebral mass of 10x8x7cm extending into vertebral canal and compressing the spinal cord. During the course of treatment Carboplatin, Etoposide, Emeset, Decadran was given.

RESULTS

Macroscopic findings*:* specimen received in three containers.

Container 1: Labelled as paravertebral component had multiple grey white, grey brown soft tissue piece each measuring 2x2x0.5cm.

Container 2: Labelled as adrenal gland has multiple grey black, grey brown soft tissue piece largest measuring 3x3x1cm and smallest measuring 0.5x0.5 cm.on cut section grey white grey brown areas identified

Container 3: Labelled as intraspinal component has multiple (2) grey black, grey brown soft tissue piece largest measuring 3x2x1 cm and smallest measuring 0.5 x0.5 cm. On cut section grey brown areas

Ganglioneuroblastoma-Intermediate Type-Stroma Rich

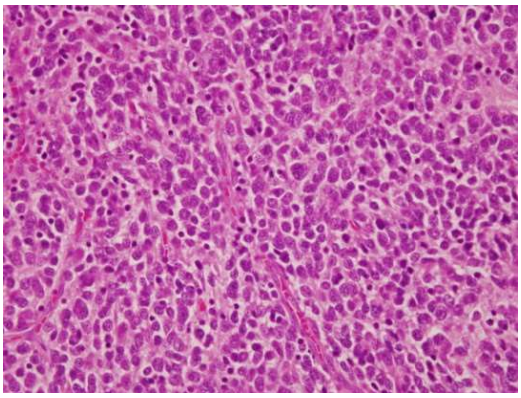


Fig. 1. Tumor cells arranged in nest separated by thin fibrous septae, H&E, 40x image

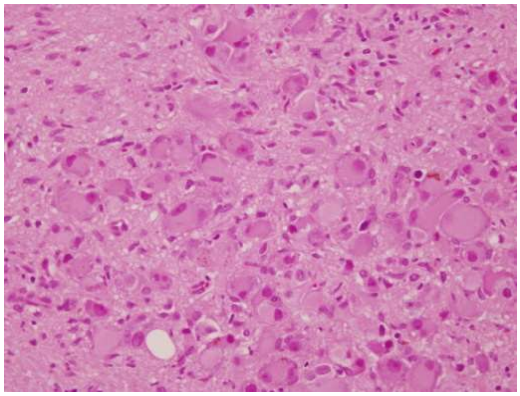


Fig. 2. Ganglion cells, H&E stain,40x

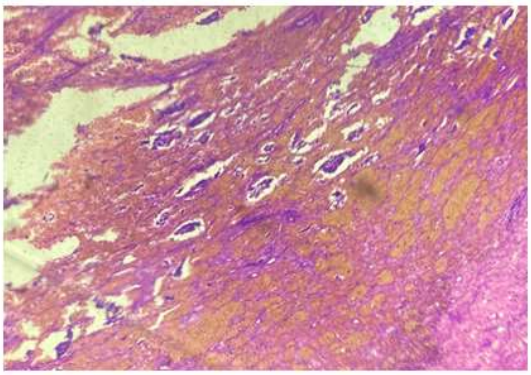
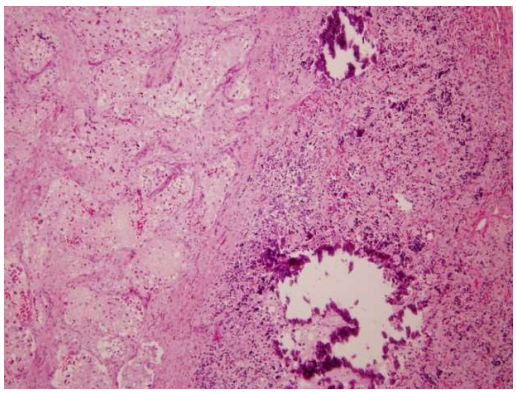


Fig. 3. Tumor cells arranged in rosette, H&E stain, 10x image



Histological examination: Multiple sections studied from container labelled as intraspinal part reveal fibrofatty and fibromuscular tissue along with a neoplastic lesion predominantly composed of variable stages of maturing ganglion cells arranged in nests and fascicles separated by schwannian matrix. Multiple sections studied from container labelled as paravertebral and adrenal gland reveal normal histology of adrenal gland and a tumor. At one foci, there are small round to oval shaped cells with hyperchromatic nuclei and scanty cytoplasm. These cells are seen in aggregates as well as in rosette. Occasional mitotic activity is present. The predominant portion of tumor shows ganglion cells in varying stage of maturation. Background shows fascicles of schwannian cells with above feature the Intermixed ganglioneuroblastoma (stroma rich) was made out.

Table 1. Immunohistochemistry Findings

Synaptophysin	Positive
S100	Positive
CD56	Positive
Chromogranin	Positive
NF	Positive
Ki 67	2-3%
TFE-3	Negative

DISCUSSION

Neuroblastic tumor arise from sympathetic nervous system and are divided into three major categories according to shimada et al: neuroblastoma, ganglioneuroblastoma and ganglioneuroma[6]. Ganglioneuroblastoma is a mixed tumor containing both immature neuroblasts and mature ganglion cells. It is a malignant tumor but less aggressive than neuroblatoma. Most of the tumors are sporadic, but in 1-2% of all cases there is a family history of neuroblastic tumors. Autosomal dominance and incomplete penetrance are characteristic of Ganglioneuroblatoma. Ganglioneuroblastoma occurs at a rate of one per 10,000 live births, making it the third most common childhood tumor. The International Neuroblastoma Pathology classification (INPC) re-defined the histological features and proposed 4 tumor categories NB (neuroblatoma) GNB-intermixed, GN (ganglioneuroma), GNB-nodular (classical). The four categories are divided in two distinct prognostic groups Favourable histology (FH) and Unfavourable histology (UH) based on the combination of age, grade of neuroblastic differentiation and mitosis – Karyorrhexis index. Staging based on international Neuroblastoma staging system. About 1% of tumor will metastasize to bone, liver, lung, brain, skin and bone marrow.[8] Diagnosis is made by ultrasonography and CT for detection of primary tumor and I^{123} MIBG for metastasis. Urinary catecholamine levels are raised especially vanily mandelate and Homovanilate.[9] Patients with N-MYC amplification or deletion of short arm of chromosome 1 have aggressive disease, regardless of age and stage of disease[3]. Amplification of N-MYC can be identified by FISH technique[10]. The therapeutic stratergy includes surgery, radiotherapy and chemotherapy.[7]

Conclusion

Ganglioneuroblastoma is typically a pediatric disease. Management of ganglioneuroblastoma needs multidisciplinary approach according to stage. The histological diagnosis and classification based on INPC predict the outcome of the tumor.[4] Surgery is the treatment of choice for stage 1 and 2, radiotherapy is used in stage 3 disease and chemotherapy is reserved for metastatic disease. Because of the rarity of the pathology, all cases should be included in the rare cancer network registry for better understanding of the disease process and management.[2]

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