

Case Report

Schwannian stroma poor calcified bilateral neuroblastoma of adrenal gland: a rare case report

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ABSTRACT

Schwannian stroma poor calcified bilateral neuroblastoma of adrenal gland in 14 year old male patient is an exceptionally uncommon, malignant tumor that is derived from the primitive neural crest cells of the adrenal medulla. Neuroblastoma is the fourth most common malignancy of childhood up to five years of age, and very occasionally seen after that. It rarely occurs in the adrenal gland after ten years of age and bilaterally. The diagnosis was established based on clinical, radiological, histopathological and immunohistochemical features. At present, the patient is on chemotherapy and radiotherapy but the recent MRI is showing bilateral pubic bone and femur metastasis. Conclusion: Schwannian stroma poor calcified bilateral neuroblastoma of adrenal gland is a rare tumor at the age of 14 years which needs precise histopathological assessment and confirmation by Immunohistochemistry.

Keywords: Schwannian stroma poor neuroblastoma, Adrenal gland, Immunohistochemistry

INTRODUCTION

Schwannian stroma, poor calcified bilateral neuroblastoma of adrenal gland in 14 year old male patient is an uncommon malignant tumour that is derived from the primitive neural crest cells of adrenal medulla.^{2,7} It is the fourth most common malignant tumour of childhood up to five year of age and rarely occurs after ten years of age.^{1,8,9} Bilateral Schwannian Stroma Poor Calcified Neuroblastoma is extremely rare tumour.^{7,8} Four types are described by International Neuroblastoma Pathology Committee (INPC)-Schwannian stroma poor neuroblastoma, Ganglioneuroblastoma intermixed, Ganglioneuroblastoma nodular, Ganglioneuroma.⁵

CASE REPORT

A 14 years old male presented with dull aching and intermittent abdominal pain, thigh pain and midline chest pain for three days, more at night. On examination- Pulse-114/min, Blood pressure-108/74 mmHg, Respiratory rate- 18/min. The systemic examination was

unremarkable. Complete blood cell count and peripheral smear examination was normal. ESR was 128 mm/1st hour. Urine examination was unremarkable. CRP-24mg/lit, S. Creatinine-0.38 mg/dl, S. Alkaline Phosphatase-89U/L, S.LDL- 812U/L. USG- Large well defined, bilateral, retroperitoneal, calcified mass, possibly adrenal lesion. CECT- Large, well defined heterogeneously enhancing soft tissue density mass lesion in the left and right retroperitoneum with multiple linear and punctate areas of scattered calcification and central hypodense areas of necrosis within its limits, most likely an adrenal lesion (Figure 1). Enlarged retroperitoneal lymph nodes are also seen.

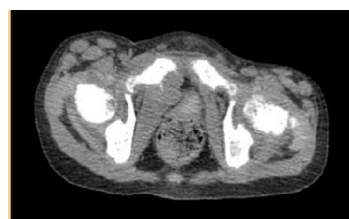


Figure 1: CECT.

Histopathology- Gross: Specimen of left radical nephrectomy along with supra-renal mass measuring 14.0x11.5x5.0 cm, weighing 400gms. The adrenal gland tumour measured 11.0x7.0x5.0 cm, was well-encapsulated, round and brown in colour. The cut surface was variegated in appearance with yellowish at center with patchy and pinpoint calcifications with areas of hemorrhage and necrosis (Figure 2). Total 42 lymph nodes were received.



Figure 2: Gross of neuroblastoma.

Microscopic examination: Tumour is composed of neuroblastic cells arranged in well formed lobules in fibrillary matrix separated by fibrovascular septa (Figure 3). Tumour cell have rim of cytoplasm & indistinct cytoplasmic borders (Figure 5). Giant & pleomorphic neuroblasts are seen. The nuclei are round to oval with coarse granular chromatin and indistinct nucleoli. Schwannian stroma formation and ganglionic differentiation is seen less than 5% of the tumour. Mitosis-karyorrhexis index is < 200/5000 cells. Mitotic figures are < 10/10 HPF. Extensive areas of haemorrhage, necrosis and intratumoural calcification are seen (Figure 4). The biopsy from right sided adrenal gland is also seen neuroblastic features (Figure 6). Tumour is reaching up to the capsule. Total 42 lymph nodes were studied from hilar, para-aortic and aortocaval areas out of which 38 lymph nodes positive for metastasis with 9 of them showing extra- nodal extension.

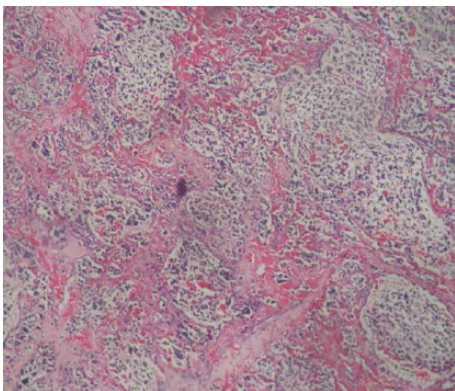


Figure 3: H&E stain (10x) Neuropils & fibrovascular septa.

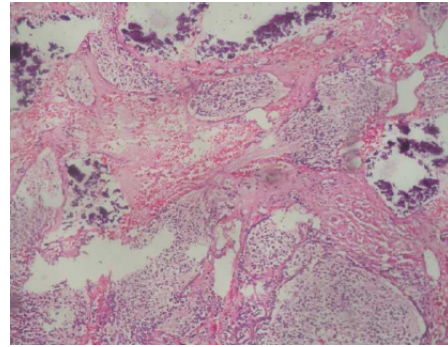


Figure 4: H&E stain (10x) Calcification.

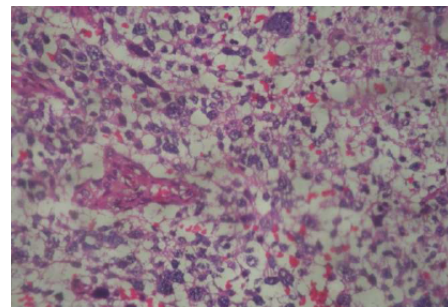


Figure 5: H&E stain (40x) Neuropils and tumour cell.

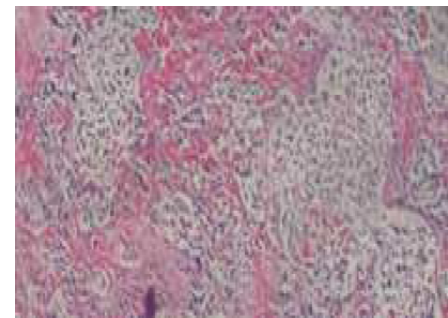


Figure 6: H&E stain (40x) right sided neuroblastoma.

Immunohistochemistry (IHC) - The tumour cells express Synaptophysin. They are immuno-negative for Desmin, Mic-2, S-100 protein, suggestive of Neuroblastoma (Figure 7).

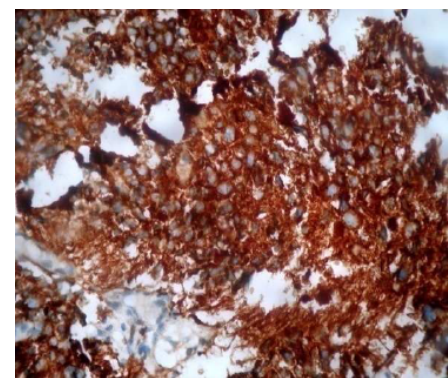


Figure 7: Synaptophysin positive.

All these findings suggestive of Schwannian stroma poor neuroblastic tumour of left adrenal gland, International Neuroblastoma Staging system, Stage: IIB.

At present patient is on chemo-radio therapy & on follow up, recent MRI is showing bilateral pubic and femoral metastasis (Figure 8).

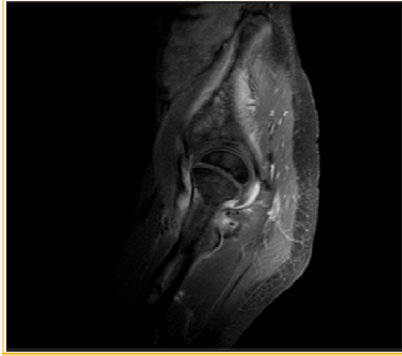


Figure 8: MRI.

DISCUSSION

Neuroblastoma of adrenal gland is rare in children after ten year of age and bilaterally.^{2,7,8,9} In neuroblastoma tumour cells are forming groups or nests separated by delicate stromal septa with limited Schwannian proliferation⁵. Neuroblastoma is classified into three sub groups – undifferentiated, poorly differentiated, differentiating.^{3,4,6} Schwannian stroma poor neuroblastoma defined as a tumour with background of readily recognizable neuropils. Most of the tumour cells in this subtype are undifferentiated and only 5% or less of the cell population has cytomorphologic features of differentiation.^{3,4,6} Supplementary techniques are required for the confirmation of the diagnosis like IHC, electron-microscopy and cytogenetics.⁶ The tumor is positive for Synaptophysin, Chromogranin, Vimentin, NB 84, Neurofilament and negative for Desmin, Mic-2 and S-100.^{6,7} The neuroblastoma is diagnosed by combination of CECT, MRI, histopathology and IHC.

REFERENCES

1. Ross JA, Severson RK, Pollock BH, et al. Childhood cancer in the United States: a geographical analysis of cases from the Pediatric Cooperative Clinical Trials Group. *Cancer* 1996; 77: 201-207.
2. MacKay B, Luna MA, Butler JJ. Adult neuroblastoma. *Cancer* 1976; 37: 1334-1351.
3. Russell DS, Rubinstein LJ. *Pathology of Tumors of the Nervous System*. Baltimore, MD: Williams & Wilkins, 1971.
4. Shimada H, Ambros IM, Dehner LP, et al. The International Neuroblastoma Pathology Classification (the Shimada System). *Cancer* 1999; 86: 364-372.
5. Shimada H, Ambros IM, Dehner LP, et al. Terminology and morphologic criteria of neuroblastic tumors. Recommendations by the International neuroblastoma Pathology Committee. *Cancer* 1999; 86:349-363.
6. Molenaar WM, Baker DL, Pleasure D, et al. The neuroendocrine and neural profiles of neuroblastoma, ganglioneuroblastomas and ganglioneuromas. *Am J Pathol* 1990; 136: 375-382.
7. Suzuki H, Honzumi M, Funada M, Tomiyama H: Metachronous bilateral adrenal neuroblastoma *Cancer* 1985; 56: 1490-1492.
8. Allan S G, Cornbleet M A, Carmichael J, Arnott S J, Smyth J F: Adult Neuroblastoma. Report of three cases and review of the literature. *Cancer* 1986; 57: 2419-2421.
9. Kaye J A, Warhol M J, Kretschmar C, Landsberg L, Frei III E: Neuroblastoma in adults. Three case reports and a review of the literature. *Cancer* 1986; 58: 1149-1157.

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