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Pathology

WILMS TUMOUR WITH RHABDOID DIFFERENTIATION A CASE REPORT

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ABSTRACT Wilms tumour is most common pediatric renal neoplasm having a peak incidence between 2-5 years of age. The classic Wilms tumour contain blastemal, epithelial and stromal component. It may differentiate into more mature mesenchymal tissue type such as skeletal muscle following chemotherapy. We present a case of 5 year female presented with history of abdominal mass for 6 month and haematuria for 2 months. CECT reveals a mass of size (13.4 x 9.8) cm^2 in abdominopelvic cavity with necrotic area. Radical nephrectomy was done following neoadjuvant chemotherapy. The histopathological examination of tumour specimen showed admixture of blastemal, tubular and mesenchymal component with extensive rhabdoid differentiation. Capsular invasion not seen. Immunohistochemisry showed positive for desmin. Chemotherapy is extremely effective in Wilms tumour but rhabdomyoblastic stromal components are more chemoresistant than the other component.

KEYWORDS : Wilms tumour, Rhabdoid differentiation, Desmin, Immunohistochemistry

INTRODUCTION

Wilms tumour is most common pediatric renal neoplasm(Mills, Carter, Greenson, Reuter, & Stoler, 2012).Peak incidence is in between 2 and 5 years of age with 90% being diagnosed by age of 6 years(Mills et al., 2012). There is no appreciable sex predilection(Rosai, 2011). It has been sparsely reported that Wilms tumour may undergo differentiation to mature appearing skeletal muscle following chemotherapy and radiation(Seifert et al., 2012). The frequency of this event is unknown(Seifert et al., 2012).

CASE REPORT

A 5 years old female presented with history of abdominal mass for 6 months and haematuria for 2 months. CECT revealed heterogeneously enhancing lobulated mass of size (13.4 x 9.8) cm^2 in abdomino pelvic cavity with necrotic areas and few calcification foci within. Truecut biopsy taken from right renal mass shows small round cells in a loose fibromyxoid stroma suggestive of small round cell tumour with consideration of possibility of Wilms tumour. The patient had received neo-adjuvant chemotherapy, CECT following chemotherapy revealed reduction in size of the mass to (10.4 x 8) cm^2. The patient underwent right sided radical nephrectomy and specimen was sent to department of Pathology, GMCH for histopathological examination. On gross examination of the specimen a tumour mass noted measuring (10.5 x 8 x 7) cm^3. Cut surface of tumour is solid, greyish white with some necrotic areas.

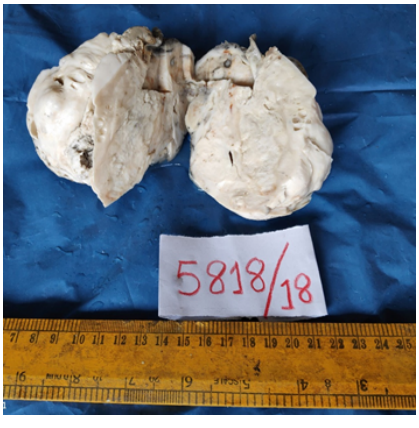

Fig 1: Grossly tumour is solid, greyish white.

Microscopic examination showed admixture of blastemal, tubular and mesenchymal component with extensive rhabdoid differentiation along with areas of necrosis. Capsular invasion and anaplasia not seen. The regional lymphnode, ureter and vessel cut margin showed no evidence of invasion.

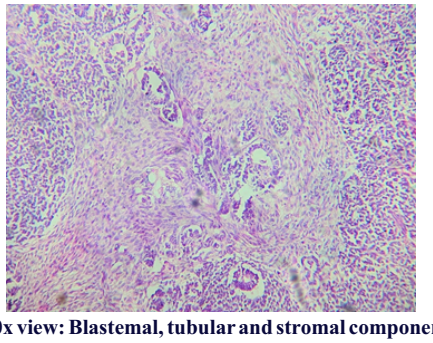

Fig 2: 100x view: Blastemal, tubular and stromal component.

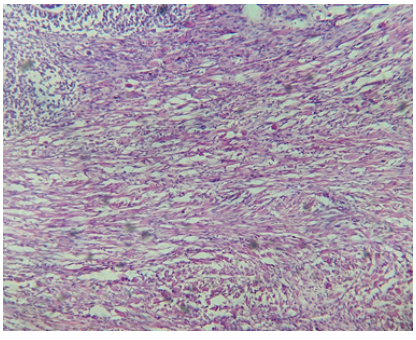

Fig 3: 100x view: Rhabdomyomatous differentiation.

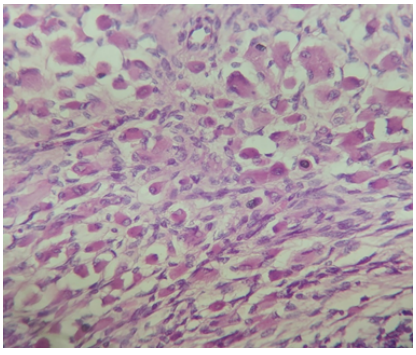

Fig 4: 400x view: Rhabdoid differentiation.

In immunohistochemical examination the mesenchymal component showed positivity for desmin in rhabdomyomatous foci.

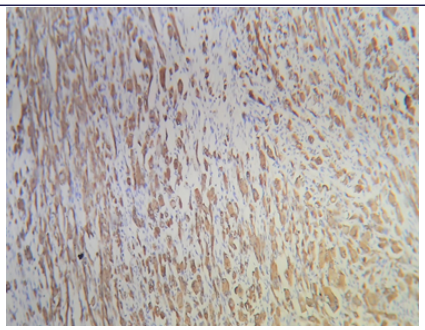

Fig 5: 400x view: Desmin positivity.

Histopathological and immunohistochemical findings revealed the diagnosis as Wilms tumour with rhabdoid differentiation .

DISCUSSION

Wilms tumour is also known as nephroblastoma, embryoma, carcinosarcoma, adenosarcoma, and adenomyosarcoma(Sebire & Vujanic, 2009). It is a neoplastic process that recapitulates embryogenesis at the morphologic and molecular levels'(Li et al., 2002). The clinical presentation of Wilms tumour is classically as abdominal mass. Pain and haematuria is rare(Rosai, 2011). Three major components, undifferentiated blastema, mesenchymal and epithelial tissue are identified in Wilms tumour(Rosai, 2011). The blastematous areas are extremely cellular and composed of small round-to-oval primitive cells; the cytoplasm is usually very scanty, but sometimes is more abundant and exhibits an oncocytoid appearance. The epithelial component is characterized by the formation of embryonic tubular structures. The mesenchymal element usually have a spindle cell fibroblast like configuration exhibit differentiation towards various cell types, particularly smooth muscle and skeletal muscle(Garvin et al., 1985). Modern chemotherapy has been extremely effective in management(Mills et al., 2012). Chemotherapy induce cell differentiation which is often rhabdomyomatous(Seifert et al., 2012). Tumour with rhabdoid differentiation is more resistant to chemotherapy and shows poor volumetric respons to pre-surgical chemotherapy. However these does not represent treatment failure or tumor aggression(Seifert et al., 2012). The chemotherapeutic agents reported to induce cell maturation include vincristine, doxorubicin, dactinomycin and dacarbazine(Seifert et al., 2012).

CONCLUSION

We report a case of Wilms tumour with rhabdoid differentiation. The rhabdoid tumour of kidney comes as differential diagnosis. Presence of distinct blastemal pattern under low power differentiate it from rhabdoid tumour of kidney. One must consider the possibility of rhabdomyomatous differentiation following neo-adjuvant chemotherapy as it may spare the patient further cycles of treatment and allow for close observation or surgical management.

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