

Immature Retroperitoneal Teratoma Presenting as Hematuria in a Newborn- An Unusual Presentation

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Abstract

We report a rare case of a large immature retroperitoneal teratoma in a newborn who presented with hematuria due to blunt trauma during delivery, which resolved spontaneously without any intervention within 3 days. The baby was noticed to have an abdominal lump since birth. His clinical examination revealed a non-tender, immobile mass occupying umbilical and iliac fossa regions. USG and MRI confirmed heterogeneous solid and cystic mass with restricted mobility measuring 7.5 cm * 7 cm * 4.4 cm extending across midline to the left and compressing aorta and IVC causing right pelviureteric obstruction. FNAC of the mass showed predominantly mature squamous cells and anucleate squamous cells along with inflammatory infiltrate. Exploratory laparotomy was done which revealed an unresectable tumour and hence only partial resection was done. The baby was started on chemotherapy histopathology of mass showed immature elements with malignant potential.

Keywords: Teratoma; Retroperitoneal Tumour

Introduction

Teratomas occur in many locations in the newborn of which the most common is the sacrococcygeal region [1]. Retroperitoneal teratomas are rare tumours which comprise 3.5 to 4% of all germ cell tumours in children [2]. Extra gonadal teratomas tend to occur in midline structures such as the anterior mediastinum, retroperitoneum, sacrococcygeal region, and pineal gland. In neonates, the risk of malignancy is greater for retroperitoneal tumors than for other sites: sacrum and coccyx, heart, neck, mediastinum, and abdomen [3]. Moreover the occurrence of an immature retroperitoneal teratoma invading the aorta and vena cava is relatively rare. Here we report a case of a newborn with retroperitoneal immature teratoma with malignant potential.

Case Report

A 3 day old male baby was admitted in NICU in view of hematuria and lump in abdomen since birth. The antenatal history was not significant. He was first born delivered normally to primi mother. A firm non-tender lump measuring approximately 7 cm * 7 cm with restricted mobility was found occupying umbilical and right hypogastric region. Systemic examination was within normal limits. Alpha fetoprotein level was 2192 ng/ml.

A plain roentgenogram of abdomen (AP and lateral) of abdomen and pelvis showed soft tissue densities with calcifications.

USG abdomen showed complex mass with solid and cystic areas with internal echoes and calcific areas measuring 7.5 cm * 7 cm * 4.4 cm medial to right kidney suggestive of cystic lesions. MRI abdomen confirmed the findings made by USG. There was no free fluid in the abdominal cavity.

Fluid aspirated showed predominantly mature squamous cells and anucleate squamous cells along with inflammatory infiltrate.

Exploratory laparotomy revealed a large mass of solid and cystic consistency in paravertebral region encasing inferior vena cava and aorta. There was no significant lymphadenopathy. As the tumour was not completely resectable due to its adherence to the major vessels, only partial excision was done.

Histopathology of mass shows tumour composed of various elements with predominantly skin and adnexal structures, lobules of adipocytes separated by fibrous septae, mature neurological tissue, lobules of immature cartilage along with focal areas showing bone, respiratory epithelium with mucosal glands, smooth muscle bundles, bone marrow with haematopoietic elements. Focally immature neuro epithelial component is seen with tumour cells arranged in rosettes, having scant cytoplasm and round hyperchromatic nucleus. The histopathologic findings were suggestive of immature retroperitoneal teratoma with malignant potential. Due to incomplete excision of the tumour, post-operative adjuvant chemotherapy was administered. The baby was started on chemotherapy with Bleomycin, Cisplatin and Etoposide.



Figure 1: Excised specimen showing adnexal structures, bone and cartilaginous elements.

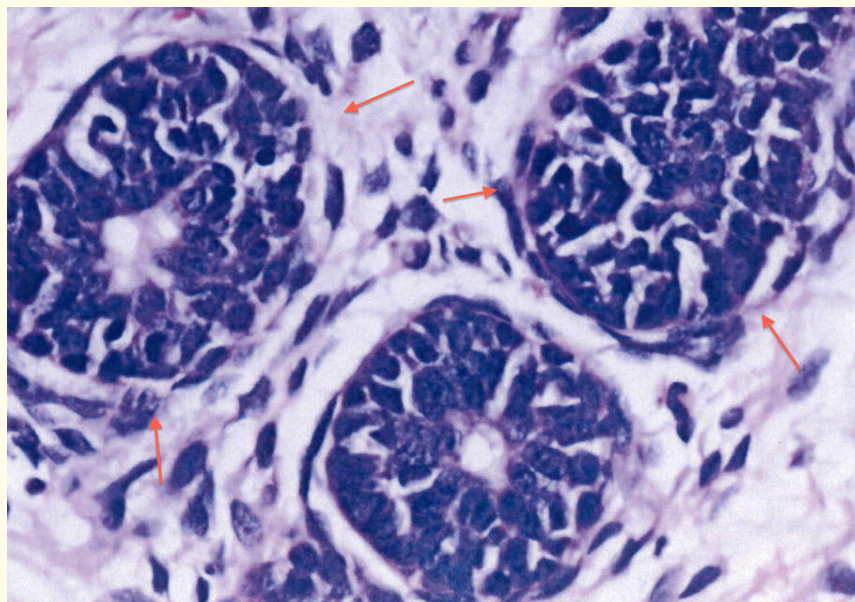


Figure 2: Histopathology slide showing tumour cells arranged in rosettes with scant cytoplasm and hyperchromatic nucleus.

Discussion

Teratomas are lesions containing elements which are derived from three primary germ layers and the most common sites are sacrococcygeal, mediastinal, retroperitoneal, and gonadal organs. Extra gonadal retroperitoneal teratomas are very rare in infants and children [4]. Retroperitoneal abdominal teratoma can present as abdominal distension, abdominal mass, hematuria and intestinal obstruction. The present case also presented with hematuria and abdominal mass [5]. The cause of hematuria was due to the rupture of the right ureter most probably due to the pressure effects of the tumour which was further compounded by extraction of the baby during delivery. Injury to the ureter is a known cause of hematuria in retroperitoneal teratomas [6]. The diagnosis of a retroperitoneal teratoma based on clinical grounds is challenging and difficult and other common conditions occurring during infancy have to be kept in mind like extra adrenal neuroblastoma, Multicystic kidney disease and Wilm's tumour. However neuroblastoma is an almost exclusively a pediatric neoplasm [7], the median age at diagnosis is 2 years and more than 90% of patients are diagnosed under ten years of age [8].

It is possible to suspect abdominal teratomas by radiological investigations: Roentgenogram, USG and CT and MRI of abdomen. Although CT is the most versatile imaging procedure for evaluation of abdominal masses, MRI can provide a better anatomic definition in several different planes [9] and has the added advantage of avoiding ionising radiation. In our case the tumour was large and encasing the inferior vena cava and aorta, so it was deemed fit to opt for an MRI. In many of the cases, USG is useful in localizing and diagnosing teratoma, but CT scan of abdomen are the most precise tools. We were also able to make a provisional diagnosis of retroperitoneal teratoma on the basis of radiological investigations. Preoperative diagnosis of teratoma may not be possible in all the cases and in these cases diagnosis has to be confirmed by histology of the excised tumour.

Complete surgical excision is the mainstay in the management of retroperitoneal teratoma. Complete tumour resection is sufficient for cure in benign teratoma. For tumours with incomplete resection, adjuvant therapy is advised with platinum based chemotherapy [10].

It is important to note that although serum alpha fetoprotein is a useful marker in retroperitoneal teratomas, it is may not be elevated preoperatively in many cases [4].

Conclusion

Most of the abdominal teratomas are benign in nature and are composed of mature cells, however 20 - 25% of these may also contain immature elements. Immature teratoma may contain variable quantities of immature neural tissues resembling embryonic components and these may co-exist along with mature tissues.

Retroperitoneal teratomas are rare in neonates but must be suspected if calcifications are found by radiological investigations. Complete surgical excision offers up to 100% survival rate, however management of tumours with incomplete excision centres around chemotherapy.

Source of Support

None.

Conflict of Interest

None.

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