



A Rare Case of Sacrococcygeal Yolk Sac Tumour

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Abstract

Extragenital yolk sac tumour is a rare variety of germ cell tumour. Here we report a case of sacrococcygeal yolk sac tumour in a 5 years old female child presenting with gluteal swelling. Although the common site of germ cell tumor in child is the gonad, however the next common site is the sacrococcygeal region.

Keywords: *yolk sac tumour (YST), gluteal region, sacrococcygeal region.*

Introduction

Sacrococcygeal germ cell tumours are the 2nd most common after gonadal germ cell tumor. These are more common in girls. Malignant germ cell tumors account for 3% of childhood neoplasm and yolk sac tumor is the most common histopathological subtype^[2].

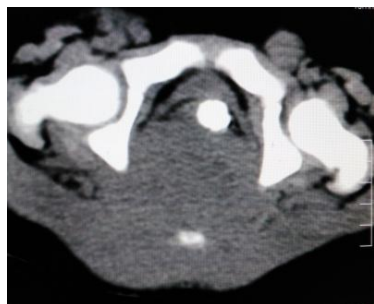
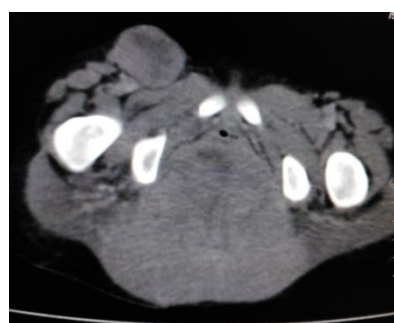
Yolk sac tumor also known as Endodermal sinus tumor (EST). This tumor may less commonly be present in "pure" form. Although, most germ cell tumors in children originate in the gonads, the most common primary site for YST is the sacrococcygeal region^[3].

Here we present a case of primary YST of the sacrococcygeal region.

Case Report

A 5 year girl was referred to the department of Radiodiagnosis for CT scan of the pelvis. The chief complaint of the patient was difficulty in micturition & defecation for the past 1 month

along with gluteal swelling. It was sudden in onset & increasing progressively. On examination of the patient swelling & thickening was noted in the gluteal region which was said to be occurred after giving an injection in the buttock. The patient had no fever & pain. CT scan revealed a large ill-defined slightly hyperdense soft tissue mass lesion in the sacrococcygeal region. There was no calcification in the mass lesion. Contrast study revealed heterogeneous contrast enhancement with some non enhancing necrotic areas. The lesion was extending inferiorly in the folds of the buttock as well as into the vulva anterior inferiorly. Anteriorly the rectum & bladder were pushed with indistinct margin in between them suggestive of probable infiltration. The muscles of the gluteal region as well as the iliopsoas, obturator & piriformis were not seen separately. Multiple enlarged lymph nodes were also noted in the inguinal region bilaterally.

**Fig: 1** Non Enhance Study**Fig: 2** Contrast Enhanced Study**Fig: 3** Contrast Enhanced Study**Fig: 4** showing inguinal lymphadenopathy

The bones of the sacrum & coccyx are destroyed but not so much. On imaging the differential diagnosis of germ cell tumor & rhabdomyosarcoma were given & biopsy was taken. Biopsy report revealed it as a yolk sac tumor.

Discussion

Germ cells in the developing embryo arise in the yolk sac, migrate around the hinder end of the primitive gut to the genital ridge on the posterior abdominal wall, and are finally absorbed into the developing gonads. It is suggested that during this migration, some germ cells may get left behind on the journey and come to rest at various sites along the dorsal wall of the embryo near the midline. Germ cell tumor is found in the testes, ovaries, and several extragonadal sites including the pre-sacral area and anterior mediastinum. Patients with sacrococcygeal yolk sac tumor present most often with complaints of constipation or buttock swelling^[3]. Sacrococcygeal YST develops usually in children less than 3 years of age^[1, 3-4]. These tumours are highly aggressive, harbouring the tendency for early lymphatic and haematological metastasis to distant sites^[5,6]. In our case we

found multiple enlarged lymph nodes in inguinal regions.

The imaging of sacral tumors in children has great values in identifying the position, contents and invasion. YST is often complicated with hemorrhage, necrosis and cystic degeneration. Obscure boundary between tumor and surrounding tissue, sacral invasion and metastases are signs of malignancy.

Gross examination of YSTs typically reveals a mass that is predominantly solid and is soft, white, gray, or pale yellow. Cystic degeneration as well as necrosis and hemorrhage are often present.

The treatment of malignant sacrococcygeal GCTs, such as primary yolk sac tumor is dependent on the extent of disease. Local disease is best managed surgically, while advanced tumor stages benefit best from adjuvant platinum based chemotherapy.

In conclusion, yolk sac tumors, whether gonadal or extragonadal, are highly aggressive. Combined approach of radioimaging and histopathologic examination is essential for early diagnosis of YST. Recognizing such tumors early is important because these tumors are sensitive to chemotherapy with increased patient survival.

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