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Case Report

Report of testicular tumour in a toddler: management beyond the testis

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***Abstract*:** Testicular tumours in prepubertal children tend to be pure yolk sac tumours detected in stage I and have good prognosis. We describe a case of a 2-year old male child with a mixed testicular tumour presenting with stage IIC disease and managed with retroperitoneal lymph node dissection for residual retroperitoneal disease post adjuvant chemotherapy.

Keywords: dissection, lymph node, mixed, retroperitoneal, testicular, tumour

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Introduction

Testicular tumours account for about 1% of paediatric solid tumours.[1] The most common pathological diagnosis in children with testicular mass is a Non-Seminomatous Germ Cell Tumour (NSGCT) of which Yolk Sac Tumour is the commonest followed by teratoma.[2] Seminoma is rare in infants; mixed germ cell tumour (GCT) is rare in the prepubertal population.[2] Herein, we present a case of 2 year old male child with NSGCT managed by retroperitoneal lymph node dissection (RPLND).

Case report

A 2-year old male child presented with a history of left testicular swelling, which was previously diagnosed as a left inguinal hernia in August 2020 at a peripheral hospital. During herniotomy at that hospital, a testicular mass (solid cystic consistency and measured 6.5 cm × 4.5 cm × 4.0 cm) was identified and the child underwent left high inguinal orchidectomy instead. The child had an uneventful recovery and underwent work-up for testicular malignancy. He was subsequently referred to our institution for expert management.

We reviewed the histopathology blocks at our institute, which suggested mixed GCT with 70% yolk sac component and 30% seminomatous component. The tumour was unifocal, and the spermatic cord margin was involved by tumour. Rete testis and lymphovascular invasion was present. The tumour markers and malignancy work-up had not been done preoperatively. Postoperatively (14th day), alpha-fetoprotein (AFP) and lactate dehydrogenase (LDH) were raised, and beta human chorionic gonadotropin (beta-HCG) was less than 2 mIU/ml. Postoperatively, computed tomography (CT) abdomen and pelvis revealed a 7.4 cm × 7.0 cm × 8.0 cm heterogeneous lesion present predominantly on the left side of the retroperitoneum, with a centrally necrotic mass effect on the left proximal ureter causing hydroureteronephrosis and displacing the left kidney posteriorly. Another 4.3 cm × 4.0 cm lesion was found to be present in the left inguinal region. Positron emission tomography (PET) scan revealed a ^{18}F-fluorodeoxyglucose (FDG)-avid 8.1 cm × 7.3 cm × 12.2 cm (SUVmax 9.4) soft tissue density lesion in the left hypochondrium and left lumber quadrants with central hypermetabolism. Left hydroureteronephrosis was present.

The child received four cycles of platinum-based chemotherapy. The serum tumour markers were assessed before each cycle, and PET-CT was done after the second and fourth cycle. The values of tumour markers have been depicted in Table 1. PET-CT after the second cycle

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of chemotherapy suggested a decrease in the left retroperitoneal mass size (now 3.4 cm × 3.2 cm × 4.1 cm) and also resolution of left hydroureteronephrosis. PET-CT after the fourth cycle of chemotherapy suggested a retroperitoneal mass of size 2.2 cm × 2.3 cm × 3.6 cm size mainly present in the left para-aortic and left lumbar region. Figure 1 shows the comparison of the three CT images.

In view of the residual retroperitoneal mass being more than 1 cm and the complete normalization of AFP levels, it was decided to proceed with RPLND and the child underwent exploratory laparotomy and left template removal after placing a double-J (DJ) stent in the left ureter cystoscopically (Figure 2). A drain was kept in the pelvis to allow for lymphatic drainage, in case lymphorrhea occured postoperatively.

Postoperatively, the child was started on feeds on the third postoperative day (POD) and a complete diet was resumed by the fifth POD. Initially, the drain output was 200 ml/day until the third POD, after which it reduced gradually and became minimal by the seventh POD and was removed. The patient was discharged on POD 14 after removing all stitches and DJ stent.

The histopathology suggested the presence of only necrotic tissue with no evidence of any malignant cells. The patient has been on follow-up for the last 2 years and is asymptomatic. He is under strict monitoring; his AFP levels are checked every 3 months and a PET scan is performed annually.

Table 1. Values of alpha-fetoprotein levels after each cycle of chemotherapy.

Chemotherapy cycle	AFP levels after the cycle (mIU)
First	7096
Second	717
Third	6
Fourth	5.7

AFP, alpha-fetoprotein.

Discussion

Testicular Yolk Sac Tumour (TYST) has bimodal age distribution and is most common in young

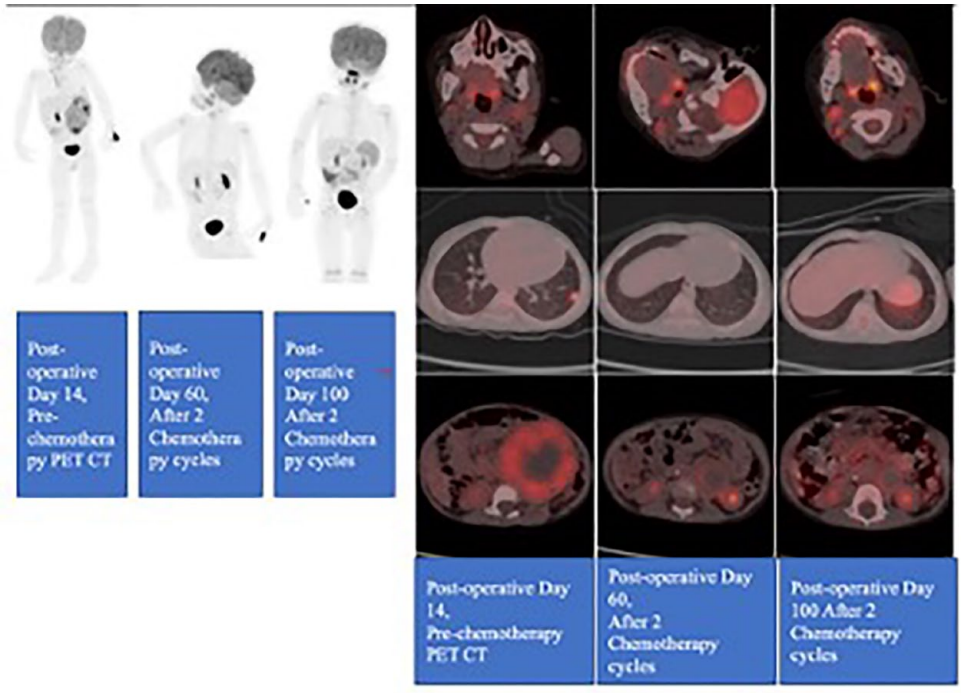

Figure 1. Comparison of the three PET-CT images with respect to the size of retroperitoneal mass.

children less than 3 years and postpubertal adults; the median age of diagnosis is 18 months.[3] Also known as infantile embryonal carcinoma, TYST is the most common testicular tumour in toddlers. The available literature has scattered case reports.[1] Infants and young children generally have pure yolk sac tumours and have a good prognosis.[4] On the contrary, yolk sac tumours in adults are often mixed and found in combination with other types of GCTs like teratoma, seminoma, choriocarcinoma and embryonal carcinoma (mixed NSGCT).[5]

Diagnosis of yolk sac tumours is based on history, clinical examination, imaging studies and blood chemistry. The usual clinical presentation is with painless solid testicular mass. Misdiagnosis is common, as was the case in the present child. He was misdiagnosed as having congenital inguinal hernia and underwent high inguinal orchiectomy without serum tumour markers. It is recommended that parents of children with risk factors for testicular malignancy such as a history of cryptorchidism, microlithiasis and a family history of cancer should be particularly vigilant.[1] Differential diagnosis includes inguinal hernia, hydrocoele and testicular inflammation.[1]

Initial presentation with metastasis is not common and occurs in less than one-tenth of cases.[1] On imaging studies [CT and magnetic resonance imaging (MRI)], yolk sac tumours present as an enhancing large solid cystic mass with intratumoural haemorrhage.[6] AFP is specific in the yolk sac tumour, but not sensitive (overall sensitivity as low as 60%).[7]

Untreated, yolk sac tumours can be aggressive and fatal. The management depends on stage of cancer and age of patient at presentation; most children present with stage I disease and have a good prognosis. Orchiectomy and platinum-based three drug chemotherapy (cisplatin, bleomycin and etoposide) with or without dissection of retroperitoneal lymph nodes is suggested. Careful postorchiectomy monitoring of serum AFP values is indicated. Most paediatric patients with metastasis or recurrence are treated with a chemotherapy regimen successfully.[8]

In stage I TYST, there is no metastasis or lymph node involvement. Therefore, the management is by high inguinal orchidectomy followed by platinum-based chemotherapy and monitoring of AFP levels. Recently, testicular-sparing surgery has been recommended for tumour size less than 2 cm. Some evidence even suggests that systematic chemotherapy may be omitted for stage I patients aged less than 1 year with no recurrence or metastasis and serum AFP stabilizing at normal levels.[1,5,6] They argue that it has no therapeutic advantage over high inguinal orchidectomy.[1,5,6]

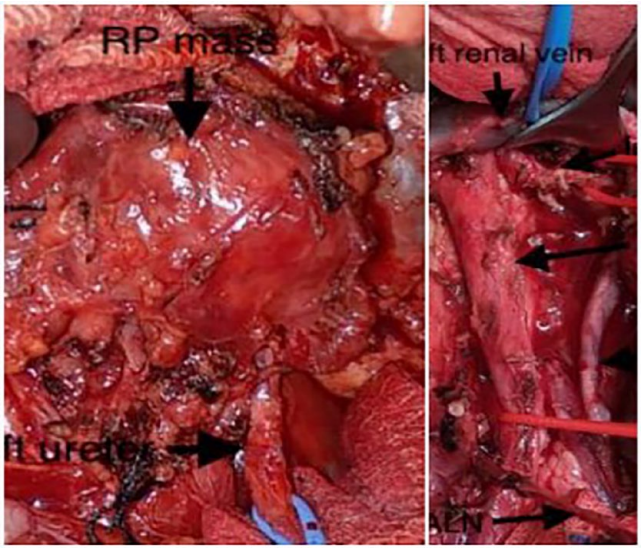

Figure 2. Intraoperative image showing the retroperitoneal mass encasing the left ureter and after removal of the retroperitoneal mass.

The role of PLND in stage I disease is controversial and not advocated in view of the following facts – metastasis in patients with TYSTs is rare and it is still unclear whether lymphangiectasis results in metastasis.[1] The associated extensive surgical dissection might lead to lymphatic fistula, enteroplegia, pulmonary atelectasis and ejaculation incompetence in later life.[1] As the therapeutic effect of chemotherapy with or without lymph node, dissection is almost same; RPLND has not been shown to provide any survival benefit in children with stage I disease.[1]

In stage II TYST, there is involvement of retroperitoneal lymph nodes, and this stage is divided into 2A, 2B and 2C depending on number and size of lymph nodes involved. RPLND or chemotherapy can be used as initial treatment after high inguinal orchidectomy in presence of detectable retroperitoneal disease. Both are effective in eradicating smaller-volume disease. After initial chemotherapy, a complete response requiring no adjuvant surgical intervention is seen in 40–71%.[6] Presence of teratomatous elements in

tumour and larger tumour masses predict failure of chemotherapy to completely resolve the retroperitoneal disease. Careful application of these two treatment modalities can produce similar survival results regardless of the sequence used. Initial use of chemotherapy usually eradicates disease in less than half of patients with stage IIA–B disease, requiring adjuvant RPLND. Finally, in selected cases where tumour markers fail to normalize, excision of the residual retroperitoneal mass may improve survival potential.[3]

The present case is unique in multiple ways. The patient had no preoperative staging before orchidectomy as he was diagnosed intraoperatively. Also, postorchidectomy work-up suggested stage IIC disease and large retroperitoneal masses for which he received four cycles of adjuvant chemotherapy. At the end of four cycles of the three drug chemotherapy – cisplatin, bleomycin and etoposide, the tumour markers normalized and the retroperitoneal mass localized and reduced in size. Hence, he was managed with RPLND.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Written informed consent for publication was obtained from the parents of the child.

Author contributions

Amit Sharma: Conceptualization; Methodology; Project administration; Resources; Writing – original draft; Writing – review & editing.

Deepak Biswal: Methodology; Writing – review & editing.

Satyadeo Sharma: Methodology; Writing – review & editing.

Kishore Roy: Methodology; Project administration; Writing – review & editing.

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Competing interests

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Availability of data and materials

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