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Case report

Primary extragonadal seminoma arising from the pelvis: A case report

C. Dracham [a], **A. Elangovan** [a,*], **R. Madan** [a], **N. Gupta** [b]

[a] *Department of Radiotherapy and Oncology, PGIMER, Chandigarh, India*
[b] *Department of Cytology and Gynaecological Pathology, PGIMER, Chandigarh, India*

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KEYWORDS
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Seminoma;
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Abstract
Introduction: Extragonadal germ cell tumors (EGCT) are relatively rare tumors with an incidence of 2–5% among all germ cell tumors and commonly seen in males. They appear mostly in midline locations, commonly in the retroperitoneum, mediastinum and central nervous system but the origin of EGCTs remain controversial.
Observation: In this case report, we present a young male with prior history of right orchidectomy for undescended testis, presented as retroperitoneal mass arising from the true pelvis. Imaging studies revealed a large mass arising in the pre-sacral region within the pelvis. Fine Needle Aspiration Cytology and Immunocytochemistry studies revealed Seminoma of classical type. The patient was treated with chemotherapy and radiotherapy, presently no evidence of disease and on routine followup.
Conclusion: This is a unique case because to the best of our knowledge, only a few cases have been reported in the literature on EGCT (seminoma in present case) presenting as a pelvic mass. Seminoma in true pelvis should be considered as a differential diagnosis when pelvic mass is detected, even in post orchidectomy cases. Tumor markers, histology, and immunohistochemistry examinations are important in making an accurate diagnosis and providing appropriate treatment.

Introduction

Germ cell tumors (GCT) are common in young males and they predominantly arise in the testis. Seminoma represents 50% of the GCTs in males. However, 2–5% of GCTs in males are extragonadal in origin and they commonly appear along the midline of the trunk [1]. The tumor can occur anywhere from the pineal gland

* Corresponding author at: Department of Radiotherapy & Oncology, PGIMER, Chandigarh, 160012, India.
E-mail address: arune195@gmail.com (A. Elangovan).

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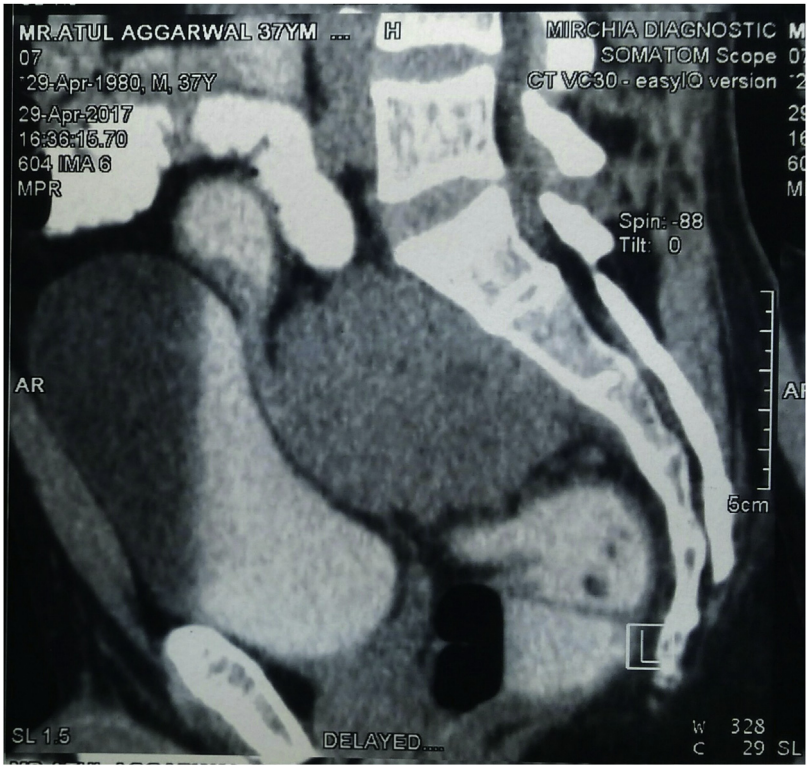

Figure 1 Contrast-Enhanced Computed Tomogram (CECT) sagittal section: showing homogenous contrast-enhanced soft tissue mass with internal calcifications in the pre-sacral region abutting the sacrum.

to the coccyx and the most common location being the retroperitoneum, followed by the mediastinum [2,3]. In this case report, we present a 35-year adult male diagnosed to have primary extragonadal seminoma arising from the true pelvis.

Case report

A 36-year old male, belongs to Caucasoid (Indo-Aryan) admixture race, presented with insidious onset low backache and pain during defecation. The symptoms were gradually progressive over six months. There was no history of upper abdominal pain, fever, jaundice, blood in stool or urinary symptoms. Patient gave the history of surgery for undescended testes on both sides and right side orchidectomy when he was five years old, however, no reports of the same were available. Physical examination revealed a well nourished young male whose chest and abdominal examinations were normal. The right testis was absent. The left spermatic cord and testis were normal on clinical examination. Incisional scars were present on both sides of groin, representative of the surgery done during his childhood.

Ultrasonogram (USG) revealed the absence of right testis. Left testis, kidneys, spleen and liver were normal. Laboratory studies revealed normal tumor marker levels α-fetoprotein (AFP: 1.51IU/ml), β-human chorionic gonadotropin (β-HCG: <0.5 ng/ml), lactate dehydrogenase (LDH:242U/L), carcino embryonic antigen (CEA: 2.8 ng/ml), normal blood cell counts and biochemistry profile. The chest X-ray and contrast enhanced computed tomography (CECT) chest were normal. CECT abdomen and pelvis showed a well marginated soft tissue mass measuring 7.3 × 7 x 6 cm in the pre-sacral region abutting the sacrum and sigmoid colon, with mild homogenous contrast enhancement and internal calcifications. There was no definite bony erosion. Similar findings were also confirmed with magnetic resonance imaging (MRI) lumbo-sacral spine. There was no mesenteric and retroperitoneal lymphadenopathy. (Fig. 1) Colonoscopy revealed extrinsic compression at 20 cm from the anal verge, however, there was no mucosal abnormality.

Cytological examination of the specimen obtained by USG guided FNAC (fine needle aspiration cytology) showed clusters of tumor cells, which were mild to moderately pleomorphic with coarse chromatin, prominent nucleoli and scant to moderate amount of vacuolated cytoplasm. On immunocytochemistry, the tumor cells were strongly positive for PLAP and negative for LCA, CD30 and pan CK. OCT4 and SALL4 immunostains are also negative. Thus overall features were of GCT, consistent with seminoma (Figs. 2 and 3).

The final diagnosis of primary retroperitoneal extragonadal seminoma arising from the true pelvis was made. We started the patient on systemic chemotherapy with BEP regimen (Bleomycin 30 IU on day1, Etoposide 100 mg/m^2 on days1–5 and Cisplatin 20 mg/m^2 on days1-5) with three weekly cycles. After four cycles of chemotherapy, the disease status was assessed with CECT abdomen and pelvis. It showed an ill-defined heterogeneous soft tissue mass lesion measuring 3 × 2.1 cm in the pre-sacral region at the S1–S2 level, suggestive of significant clinical response with a small residual tumor. Then patient received radical radiation therapy (36 Gy/20 fractions/4 weeks) by 3-dimensional conformal radiotherapy (3DCRT) technique. There was no evidence of residual disease on the follow-up imaging studies. Now patient is alive after one year post treatment, without signs of local recurrence and metastasis. Informed consent was obtained from the patient for reporting purpose.

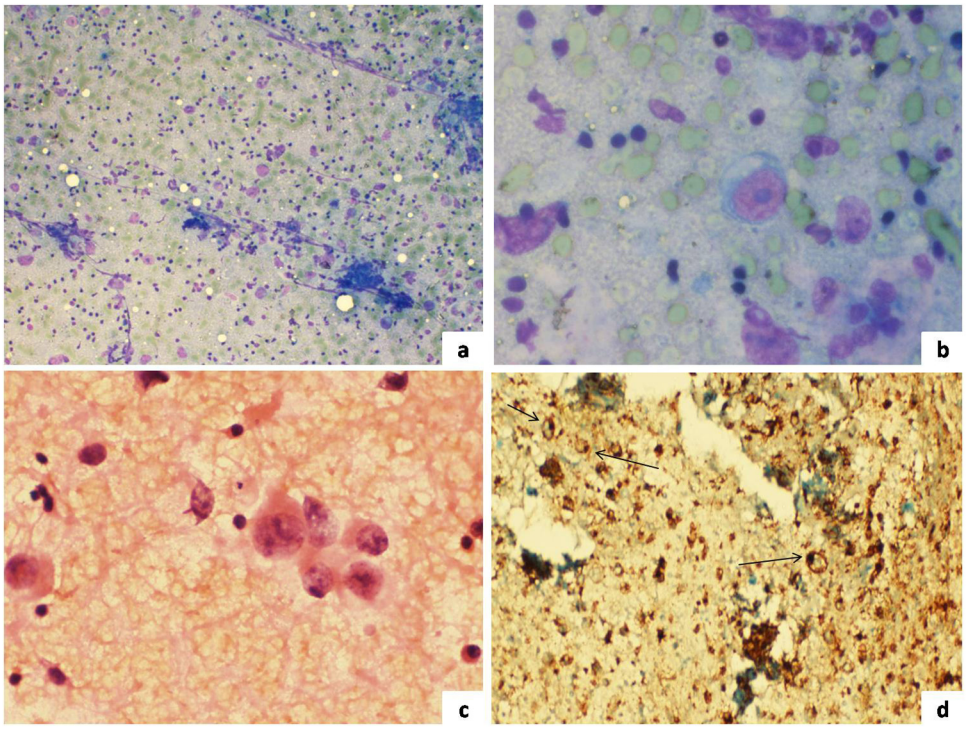

Figure 2 (a) Smear showing scattered large atypical cells admixed with lymphocytes (May-Grünwald Giemsa, 100×). (b) Smear showing large atypical cells with round nuclei, coarse chromatin and prominent nucleoli with naked atypical cells and lymphocytes in the background (May-Grünwald Giemsa, 400×). (c) Smear showing cluster of similar atypical cells (Hematoxylin and eosin, 400×). (d) Atypical cells showing cytoplasmic positivity for PLAP (ICC on cell block for PLAP, 100×, arrows pointing the positive atypical cells).

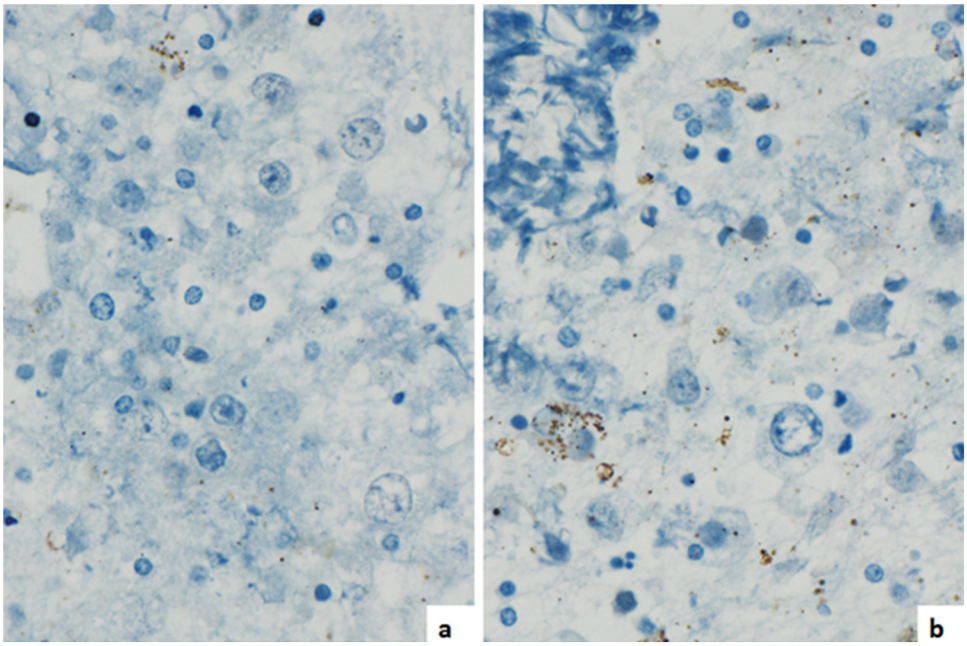

Figure 3 (a,b) Atypical cells are negative for SALL4 and OCT4 (ICC on cell block for SALL4 and OCT4, 400X).

Discussion

Testicular cancers are the most frequent solid neoplasms in young males aged 20–35 years [4,5]. The incidence of testicular cancers is greater in cryptorchidic testicles and patients with personal history of a tumor in contralateral testis. Trauma, gonadal dysgenesis, and testicular atrophy have been proposed as other etiological factors for testicular tumors [6]. In general population, the incidence of cryptorchidism is around 2%. Undescended testicles are most palpable in the inguinal canal or remain in the abdomen [7]. Abdominal tes-

ticles have a greater chance of developing cancer because of higher temperature. Winter C and Albers P [8] found that 30% of abdominal testicles have a chance of turning malignant and 90% of those tumors were reported to be pure seminomas [9,10]. Orchidopexy does not eliminate the cancer risk but early orchidopexy allows early diagnosis and treatment because of testicle being accessible for clinical examination. Two cases of extragonadal primary GCT (immature teratoma and seminoma) in the pelvis have been reported in the literature [11,12]. We have reported one such rare case in this article.

The exact origin of extragonadal GCTs (EGCT) is not known, but there are several hypotheses relating to EGCTs histogenesis. They can arise from germ cell migration along the urogenital ridge during embryonal development [13], or due to physiological disruption of germ cells in the development of regulatory functions and the delivery of genetic information to somatic sites [14]. Another theory put forward is the theory of 'burned out testicular primary'. This theory states that germ cell tumors arising in the testis have vanished after metastasizing to retroperitoneum and mediastinum [15]. Other reported rare sites are an orbit, palate, supra-sellar area, thyroid, submandibular region, liver, stomach, anterior abdominal wall, vagina, pelvis, and prostate [16]. Prognosis of these patients depends on the histology and the initial tumor stage.

Standard treatment for EGCTs remains controversial because of the rarity of the disease. Serum tumor markers can guide towards the diagnosis and treatment sequence during the management of EGCTs. Chemotherapy followed by surgery for removal of the residual disease is indicated in cases presenting with elevated tumor markers. Chemotherapy followed by radiation therapy is an option for remaining cases with normal limits of tumor markers. However systemic chemotherapy remains the nodal treatment for EGCTs [17,18]. Our patient received four cycles of chemotherapy followed by External beam Radiotherapy to the residual disease.

Conclusion

Primary seminoma in the true pelvis is a rare condition but should be considered in the differential diagnosis when pelvic or abdominal retroperitoneal mass is detected, even in post orchidectomy cases. This is due to the present day practice of orchidopexy in childhood and orchidectomy in the post-adolescent age. Thorough testicular examination, tumor markers, and ultrasonography are mandatory in all retroperitoneal tumors. Histology and immunohistochemistry examinations are important in making an accurate diagnosis and providing appropriate treatment. Multimodality treatment (surgery, chemotherapy and radiation therapy) is recommended depending on histopathological features. Most of the patients can be cured if they diagnosed prior to visceral metastatic dissemination.

Conflict of interests

None.

Consent from the patient

Obtained.

Authors' contributions

All the authors critically reviewed the manuscript for its content, contributed to the interpretation and presentation of the review, and approved the final version of the same before submission.

Specific contributions by the authors individually has been highlighted below:

1. Dr. Chinnababu Dracham — Constructed the idea for case report; Prepared the manuscript, organised and supervised the course of the article.
2. Dr. Arun Elangovan — Responsible for the patient's management, follow up.
3. Dr. Renu Madan — Critically reviewed the article before submission not only for spelling and grammar but also for its intellectual content.
4. Dr. Nalini Gupta — Provided the tissue diagnosis, performed the immunocytochemistry for its confirmation and delivered the images regarding the same.

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