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Mixed Germ Cell Tumor of Testes Presenting As Chest Wall Mass – An Extremely Rare Entity with Diagnostic Challenge

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Abstract
Mixed germ cell tumour is a kind of testicular tumour with more than one germ cell component, and are more commoner than the pure histological forms[1]. We present a rare case of a mixed germ cell tumour composed of embryonal carcinoma and yolk sac tumour, presented as chest wall mass. This case is being reported here due to dilemmatic way of presentation and also to emphasize that histopathology can be misleading and immunohistochemistry is necessary in such cases.

Keywords*: Testicular malignancy, mixed germ cell tumour, chest wall metastasis, orchidectomy.*

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I. Introduction

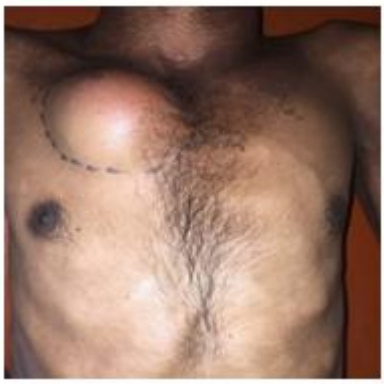
Testicular neoplasms although are very rare, accounting for 1% of the malignancies in men. These tumours are the most common solid tumours in young adult men between 20 and 40 years of age. More than 90% of testicular neoplasms correspond to germ cell tumours that are divided into seminomatous and non seminomatous tumours, corresponding to the histologic subtypes of embryonal carcinoma, yolk-sac tumour, teratoma and choriocarcinoma[7]. More than half of the germ cell tumours contain more than one cell type and are therefore known as mixed germ cell tumours and they are much more common than any of the pure histologic forms representing 32%-60% of all germ cell tumours. Essentially, any admixture of the germ cell tumours as seen in pure form may be seen, one of the most common admixtures being embryonal carcinoma and teratoma . Minor foci of yolk sac tumour are common, although it is usually overshadowed by other components, such as embryonal carcinoma. Although seminoma may be seen as part of a mixed germ cell tumour, in some cases one sees seminoma separate from a dominant mass of non-seminomatous mixed germ cell neoplasia, and in such cases it is probably truly multicentric neoplasia, although for sign-out purposes it is probably sufficient to consider the seminoma together with the other neoplastic components under the one designation of mixed germ cell tumour with the traditional rough quantitation of the various components in descending order of frequency. The average age of presentation for patients with mixed germ cell tumours is 30 years. Unfortunately, many of these patients present late, usually with some or the other complications which are difficult to treat and carry bad prognosis. Still, if they can complete the chemotherapy they have a reasonable survival period, depending on the complications they have. Although pure embryonal carcinoma is a relatively rare neoplasm, it is the most common component in mixed germ cell tumours. Embryonal carcinoma tends to occur at a young age. Many (66%) patients with a tumour composed predominantly of embryonal carcinoma, have metastasis at the time of diagnosis; subcutaneous metastasis of embryonal carcinoma is rare. We report on a patient who represents this unusual mixed variety of germ cell tumour embryonal carcinoma dominant form presented with chest wall mass .

II. Case Presentation

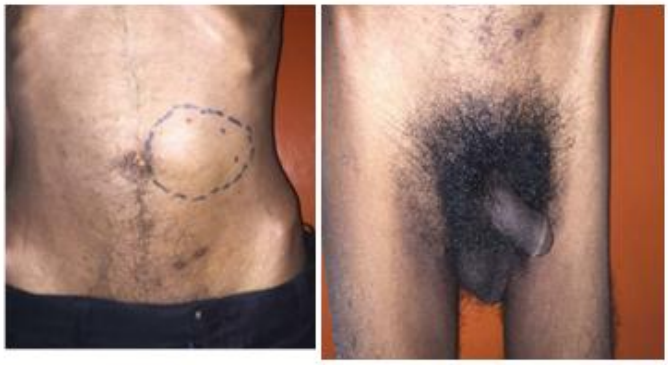
A 34 years old man presented with a gradually increasing, subcutaneous swelling in the anterior chest wall over 6months duration. The swelling measured 10 x 11 cm, was firm to hard in consistency, fixed to the chest wall, and was non tender. On other systemic examination revealed hard, not mobile lump of 6x7 cm in left lumbar region ,the patient examined in detail found with nodularity in left testis without obvious mass and left supraclavicular node size of 2*1 cm enlargement. Atypical presentation of testicular tumours are considered and the patient evaluated further with radiological and invasive investigations.

USG of the scrotum revealednodularity in left testis around 2*2 cm in upper pole.It appeared heterogeneous with irregular borders with echogenic foci of haemorrhage.

USG of the abdomen revealed multiple liver metastasis with enlargement of the retroperitoneal and para aortic nodes.



Clinical picture of a) chest wall massb) abdominal mass c) no obivious testicular mass



CT chest(figure 1) showed metastatic deposits along the right chest wall as heterogeneously enhancing mass lesion extending into thoracic cavity, anterior mediastinum with size of 9.2*11.2*11.4 cm with sternum erosion,and mediastinal lymph nodes, para tracheal ,and left supraclavicular nodes enlargement.
CT abdomen & pelvis (figure 2) showed multiple liver metastasis, heterogeneous mass lesion size of 16.7*7.5*14.1 cm noted in para aortic region with moderate free fluid in abdomen &pelvis.

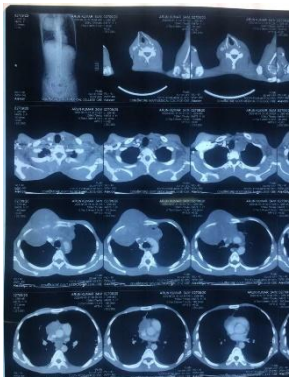


Figure 1: CT CHEST

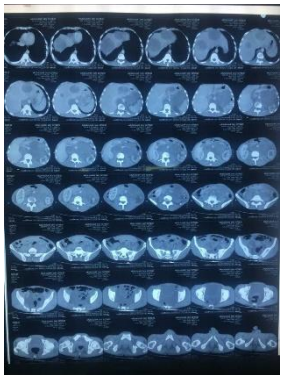
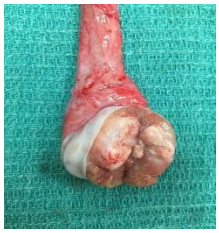


Figure 2: CT ABDOMEN

Pre biopsy Tumour markers: Alpha-Fetoprotein (AFP):60081 ng/ml, Beta-HCG:162 mIU/ml, LDH:3500 U/L
FNAC from chest wall mass-malignant cells arranged in sheets discohesive clusters and vague acinar pattern - suggestive of metastatic deposits.
Patient underwent left high inguinal orchidectomy.
On gross pathological examination, there was a, solitary, non encapsulated tumour measuring 2 x1 x1cm located upper pole of left testis. The tumour was hard in palpation. There were areas of haemorrhage and necrosis with yellowish discoloration. Histopathology showed tumour cells arranged in papillary and glandular pattern with areas of necrosis with diagnostic difficulty to differentiate it from adenocarcinoma rete testes from mixed germ cell tumor .



Clinical picture of bisected testis

Biopsy of Post op specimen revealed Tumour cells arranged in glandular and papillary pattern suggested adenocarcinoma rete testis which was later subjected to immunohistochemistry which confirmed it as mixed germ cell tumor.

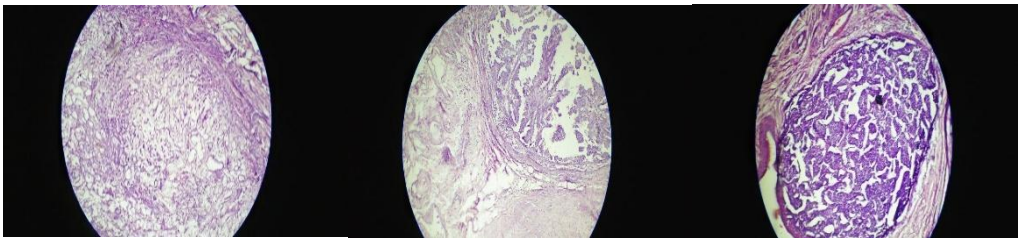


Fig 3*-yolk sac component showing reticular/ microcystic pattern with loose stroma.*

Fig 4*– embryonal component showing papillary pattern with cells exhibiting high grade features*

Fig 5 *- embryonal component*

IHC- the yolk sac component is positive for AFP; the embryonal component is positive for CD 30 and focal immunoreactivity for PLAP. CK7 highlights rete testis only.MIXED GERM CELL TUMOR (YOLK SAC COMPONENT 40%;EMBRYONAL CARCINOMA COMPONENT 60%)

Post orchidectomy tumour markers-: Alpha-Fetoprotein (AFP):92317 ng/ml, Beta-HCG:242 mIU/ml, LDH:4500 U/L

Since the patient was in stage IV disease. The patient was given chemotherapy of the BEP regimen, i.e. Bleomycin, Etoposide, and Cisplatin with Radiotherapy. The poor prognosis was explained to him and to his relatives. He was treated symptomatically. However, finally he succumbed during his early palliative care.

III. Discussion

Testicular cancer is a relatively rare neoplasm; It make up approximately two percent of all malignant cancers in men and account for up to ten percent of all malignant disease occurring within the male genitourinary system. Most of these tumours occur in three age groups - infancy, late adolescence and early adulthood. More importantly, testicular malignancy , developing in men between 20 and 40 years of age and are the third leading cause of death among men of this age group.

Pathologically, testicular cancers are divided into two classes; germ cell tumours which are derived from germinal epithelium and non-germ cell tumours which are of gonadal stroma origin. Tumours of germ cell origin comprise about 95% of all testis cancer. Germ cell tumours are divided into two basic groups: seminomas which occur in approximately 40% of the population and non-seminomatous tumours (NSGC) which may be seen in pure or mixed form.

The diagnosis of testicular cancer focuses on clinical and instrumental elements. Cryptorchidism is the most significant risk factor for testicular cancer, increasing the risk up to 11-fold. Testicular cancer classically manifests as a painless swelling or enlargement of the testicle. However, about 10% of patients manifest new-onset testicular pain. Additionally, nearly one-fourth of patients with metastatic disease complain of metastasis-related symptoms (e.g. low back pain caused by metastatic lymph nodes or a primary tumor located in the retroperitoneal space or presents as swelling in the chest wall as in our case). Scrotal ultrasound, as a first-step test, shows lesions that are dimensionally higher than 5 mm and allows the discernment of cystic lesions from neoplastic ones, while also taking vascularization into account. The serum markers α-fetoprotein, β-human chorionic gonadotropin, and lactate dehydrogenase can be useful for diagnosis, treatment, and surveillance [4] . CT images show the presence of an intratesticular mass that is often morphologically patchy, containing ill-defined necrotic spots. CT also allows to pinpoint potential secondary tumours. Magnetic resonance imaging is rarely indicated because of the costs and superimposed results with regard to CT[2] . Radical orchiectomy represents the primary treatment for most patients presenting with a suspicious testicular mass. Orchiectomy is both diagnostic and therapeutic[1,9].

Given that testicular cancers are curable even in the presence of metastatic disease, the correct staging is a critical component for treatment decision-making and the prognosis. Testicular cancer typically spreads via the lymphatic system in a step-wise sequence. The primary lymphatic drainage of the testis is to the para-aortic lymph nodes at the level of the renal vessels. From there, the cancer may spread to the iliac and mediastinal lymph nodes, and then on to the neck lymph nodes. In advanced disease, haematogenous dissemination to distant organs may occur at sites including the lung, liver, brain, and bone[1,3]. The prognosis and therapy of these testicular tumours depend on clinical stage and histopathological type. Biological markers including AFP, HCG, and LDH are valuable in follow up the patients. Over half of germ cell tumours consists of more than one cell type, requiring appropriate sampling for correct diagnosis and correlate with serum tumour markers and immune histochemistry. Review of the literature shows different combinations of mixed germ cell tumours and we report a case of mixed germ cell tumour with rare combination in our institute.

The decision on the mode of therapy largely relies on pathologic classification of germ cell tumours and spread of tumour. Between 65–85% of all seminomas are clinically confined to the testis, whereas 60–70% of non-seminomas present as recognizable metastatic disease. Treatment of nonseminomatous or mixed germ cell tumours largely depends on whether the tumour is localized to the testis or has already metastasized to retroperitoneal lymph-nodes or other sites. Initial clinical and radiographic examination plays very important role in the management of the patient.

The established treatment of low-stage seminoma is inguinal orchidectomy followed by therapeutic or adjuvant radiation therapy. The optimal treatment of patients who present with distant metastases or bulky retroperitoneal disease is initial chemotherapy[10]. The role of salvage chemotherapy, surgical removal, or radiation therapy for persistent radiographic masses remains controversial. Of major importance is the fact that roughly one third of patients with histologically pure seminoma of the testis who ultimately die of the disease are found to harbour nonseminomatous elements in metastatic sites .

Patients presenting with nonseminomatous germ cell tumours are subdivided into low, high and advanced stage disease. Patients with low stage may be candidates for surveillance, chemotherapy, or retroperitoneal lymph node dissection, depending on variety of factors such as the clinical staging, serum tumour markers, and tumour histologic findings. On the other hand, patients with advanced disease are further subcategorized into good and poor risk categories and are then subjected to primary and secondary chemotherapy depending on the nature of their disease.

IV. Conclusion

A rare combination of mixed germ cell tumor with chest wall metastasis is very rare[3]. Only four cases of Cutaneous metastasis of embryonal carcinoma and choriocarcinoma noted[11].Although the overwhelming majority are malignant, they are curable.Since embryonal carcinoma component having aggressive tendency to early haematogenous spread the need for early detection and treatment is thus emphasised. Early diagnosis and treatment with platinum based chemotherapy in conjunction with radiotherapy and surgery have high cure rate

CONSENT

The authors would like to thank the patient's relatives for providing informed consent for the publication of this case report.

CONFLICT OF INTEREST

Authors have no conflict of interest to declare

AUTHOR CONTRIBUTION

1.Patient management and treatment decisions.
2. Patient management, surgical treatment and manuscript writing.
3. Patient management, manuscript writing.

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