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Research Paper

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Extragonadal Germ Cell Tumor in Inguinal Region- A Rare Case Report

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ABSTRACT

We present a rare case of primary extragonadal germ cell tumor (EGGCT) initially presenting with right inguinal mass since 6 months. The mass was resected along with testis and testis did not show any evidence of tumor grossly and microscopically. Histopathological examination of the surgical specimen showed extragonadal germ cell tumor.

Introduction-

Extragonadal germ cell tumors (EGGCT) are rare, representing 3-5% of all germ cell tumors(1) Men are affected more than women. They are characterised by their location on the midline from pineal gland to coccyx. Most common site of EGGCT is mediastinum (50-70%), followed by retroperitoneum (30-40%) the pineal gland (5%) and sacrococcygeal area (less than 5%). Other less common sites include orbit, suprasellar area, palate, thyroid, submandibular region, anterior abdominal wall, stomach, liver, vagina and prostate (2) They are distinct from testicular cancer in biological behaviour, natural history and management (3).

Rarity of this lesion at this site makes it unique and hence merits reporting.

Case report-

A 22 year old male presented in surgery OPD with symptoms of right sided inguinal pain and swelling since past six months. His contrast enhanced CT scan of abdomen and pelvis revealed large fungating irregular anterior abdominal mass in right inguinal region in subcutaneous planes expanding inferiorly involving scrotal skin and subcutaneous tissue and rectus abdominus muscle. This finding was suggestive of local infiltrative malignant lesion. Necrotic bilateral inguinal and right iliac lymphadenopathy was also noted. Examination for metastases including chest X-ray, chest CT demonstrated no abnormalities. His past medical history was unremarkable and had no urologic problems. His haematological and biochemical investigations were within normal limits. Right partial orchidectomy with mass was resected and sent for histopathological examination.

Gross

1. Skin covered mass along with attached right testis. Mass measuring 23x9x8cms. Cut Surface- grey white, friable, with areas of hemorrhage & necrosis.
2. Testis measuring 6x5x3cms. Cut Surface- unremarkable.
3. One inguinal lymph node was identified in the mass.

Microscopy

1. Sections from the mass revealed highly malignant tumor cells arranged in lobules, sheets and papillary pattern. The individual tumor cells are highly pleomorphic with indistinct cell outlines, many of them showing clear cytoplasm with round to irregular hyperchromatic nuclei and prominent nucleoli. Plenty of hemorrhage and necrosis was seen in the background. Abundant mitotic figures seen. (Figure 2, 3, 4)
2. Sections from the testis showed normal histomorphological features. (Figure 7)
3. Sections from the lymph node showed tumor cells replacing the entire lymph node with focal remnants of lymphoid tissue. (Figure 6)

Based on the gross and microscopic findings, following differential diagnosis were considered

1) Poorly differentiated carcinoma
2) Metastatic tumor
3) Germ cell tumor
4) Malignant melanoma.

A panel of immunohistochemistry markers were performed whose results are as follows

POSITIVE MARKERS	CK+, CD30
NEGATIVE MARKERS	S100, HMB45, CD 31

IHC clinched the diagnosis of Embryonal cell carcinoma.

Figure 1: Shows skin covered mass measuring 23x9x8 cms, along with testis measuring 6x5x3 cm. Cut surface of the mass shows grey white ,friable with areas of hemorrhage and necrosis

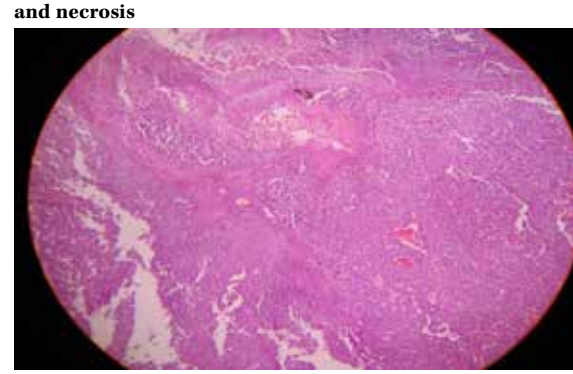

Figure 2: H &E- 4X Section from mass showing tumor cells arranged in lobules and sheets.

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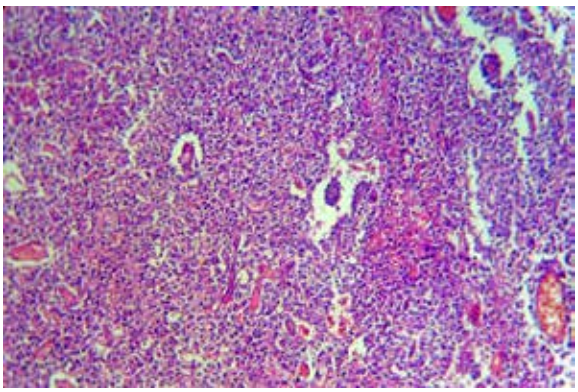

Figure 3:H &E-10 X Section from mass showing tumor cells arranged in sheets and papillary pattern. Many of them showing clear cytoplasm with areas of hemorrhage.

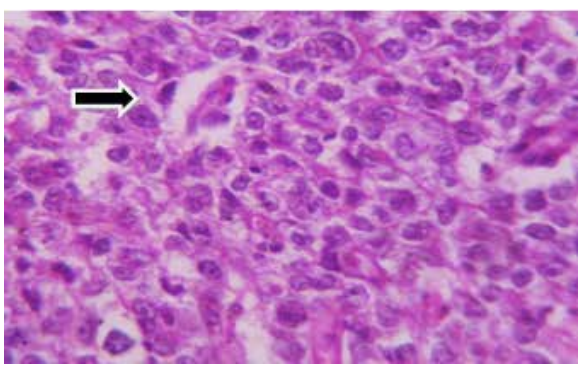

Figure 4 : H &E-40X Showing malignant tumor cells with nuclear pleomorphism, prominent nucleoli, clear cytoplasm, ill defined cell borders and plenty of mitotic figures(arrowhead).

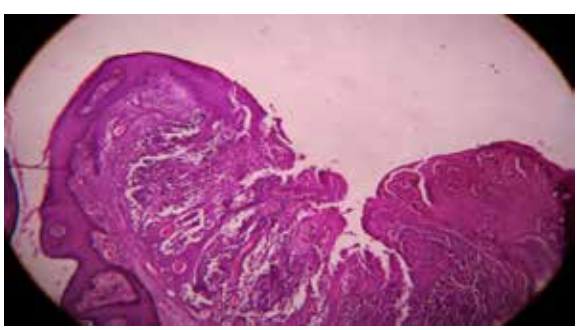

Figure 5 :H &E-4X Showing tumor cells infiltrating the epidermis.

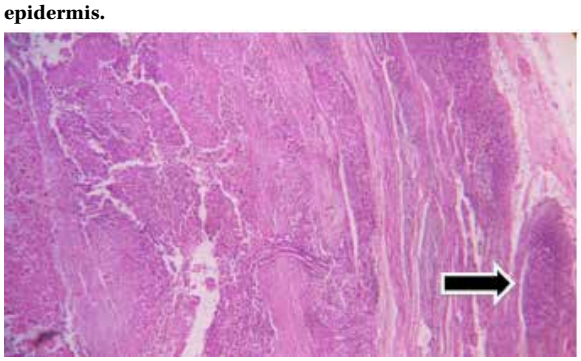

Figure 6: H&E-10X Section from lymph node showing tumor cells replacing the entire inguinal lymph node (arrow head representing remnant of lymphoid tissue)

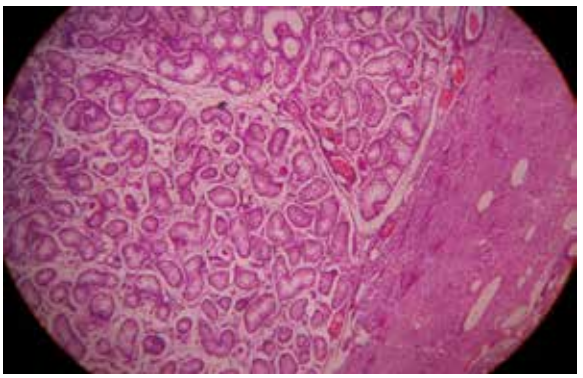

Figure 7: : H&E-10X showing normal histomorphological features of testis.

Discussion-

Primary EGGCT are rare and account a small percent, 3 to 5 % of all Germ cell tumors (GCT) (1). The majority of GCT originates from gonad, whereas a minority may occur in extragonadal places such as mediastinum, retroperitoneum and pineal gland(4). The prevalence of primary EGGCT is greater in young adults, reflecting overall higher frequency of GCT in men(4) (5). In adults, only benign EGGCT occurs at equal frequency in males and females. In children, benign and malignant EGGCT occurs equally in males and females. Exact pathogenesis of GCT is uncertain. However, according to widely accepted theory, during embryogenesis, germ cells are misplaced anywhere in the midline. Malignant transformation of these cells leads to EGGCT at these regions (2).

All GCT arise from undifferentiated malignant germ cells, which can be classified into a variety of histologic types analogous to their benign counterpart (5). Prognosis of EGGCT is worse than that of primary testicular germ cell tumors (TGCTs) due to their relative chemoresistance and advanced stage at diagnosis. EGGCT patients are at an increased risk of subsequent TGCTs and have relatively high incidence of intratubular germ cell neoplasia(6). During the past two decades these cancers have become curable neoplasm. The main reason is that these types of tumors are highly sensitive for cisplatinum based chemotherapy protocols (1).

Often patients with GCT present late, after their tumors have reached large dimensions. Presenting symptoms are abdominal mass with or without pain, backache and weight loss. Symptoms vary depending on the site and size of the tumor. Those arising in nonvital organs can reach large sizes before becoming symptomatic, but small tumors may result in significant symptoms if they obstruct, compress or rupture into important structures(2).

No abnormality can be found in the testis at initial presentation. It may take as many as 8-18 years after removal of retroperitoneal metastasis for testicular primary lesion to manifest (5).

Proposed World Health Organisation Histologic Classification of Germ cell Tumors
• Precursor lesion
Intratubular undifferentiated malignant germ cell
• Tumors of one histologic type
Seminoma
Spermatocytic seminoma
Embryonal carcinoma
Yolk sac tumor
Choriocarcinoma
Teratoma
• Mature
• Immature
• With malignant areas

• Tumors of more than one histologic type (specify each type and amount of each)
Embryonal carcinoma, Yolk sac tumor, Teratoma and syncytiotrophoblasts

Tumor markers serum alpha fetoprotein (AFP) and/ or beta subunit of human chorionic gonadotropin(beta-hCG) is produced in extragonadal non seminomatous germ cell tumors. These tumor markers provide diagnostic, staging and prognostic information. These levels are to be checked before and then at regular intervals after therapy (2). High levels are associated with poor survival (>500µgram/mL for AFP or >1,000µm/ml for HCG)(5).Serum AFP elevations are seen in yolk sac tumors. Pure seminomas and pure choriocarcinomas do not produce AFP (2). Xavier Leroy et al in their study suggested that, 11/14 Embryonal carcinomas were CD30 positive /CD117 negative (8).

Treatment and prognosis depend on the histologic type, the site of primary tumor and extent of malignant disease (7).

Conclusion-

We would like to conclude by saying that extragonadal germ cell tumors are rare and even more rare in the inguinal region. Diagnosis is supported by its characteristic features on histopathology and confirmed on immunohistochemistry. Hence, in young adults who present with inguinal swelling, a differential diagnosis of EGGCT should also be considered and careful evaluation of testis is crucial for determining whether the lesion is primary or secondary.

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