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CASE AND REVIEW OF LITERATURE**

EXTRAGONADAL MIXED GERM CELL TUMOUR OF THE FACE: REPORT OF A RARE CASE AND REVIEW OF LITERATURE

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ABSTRACT

Introduction: Germ cell tumours are a heterogenous group of tumours that could be either benign or malignant, and they primarily present as gonadal tumours. Extragenadal germ cell tumours (EGCT) are rare and represent 1-5% of all germ cell tumours and they arise in anatomical locations other than the gonads. Extragenadal germ cell tumours broadly include seminomatous and non-seminomatous tumours, with different morphologies which include mature cystic teratoma, immature teratoma, seminoma, yolk sac tumour, embryonal carcinoma, choriocarcinoma and mixed germ cell tumour. Extragenadal germ cell tumours mostly affect children and young adults, usually involve the structures along the midline of the body and are more common in males. Head and neck region is an uncommon primary site for EGCT with most tumours being benign and teratoma being the main histologic type. The exact aetiology is unknown and it has a poor prognosis compared with its gonadal counterparts.

Case: We report a case of a 3-year old girl who presented to a peripheral private facility with a right facial swelling. She had incisional biopsy done which was uneventful. The tissue was processed in our

histopathology laboratory and a diagnosis of extragenadal mixed germ cell tumour was made.

Conclusion: Extragenadal mixed germ cell tumours of the head and neck are rare and accurate histological diagnosis is essential for correct management of the patient.

Keywords: extragenadal tumour, teratoma, yolk sac

INTRODUCTION

Germ cell tumours are neoplasms that form from reproductive cells which could be either benign or malignant. Most of the germ cells tumours occur in the testicles or the ovaries and commonly affect children and young adults.¹ Extragenadal germ cell tumours (EGCTs) are rare neoplasms comprising of 1-5% of all germ cell tumours. They broadly include seminomatous and non seminomatous tumours, with different morphologies which include mature cystic teratoma, immature teratoma, seminoma, yolk sac tumour, embryonal carcinoma, choriocarcinoma and mixed germ cell tumour. EGCT composed of two or more subtypes are referred to as mixed germ cell tumours. EGCT are more common in males and mostly affects children and young adults. They arise in anatomical locations other than the gonads and favored midline locations of the body from the brain to the coccyx. The

most common anatomical locations are the mediastinum, retroperitoneum and the brain.^{2,3} Head and neck region is an extremely rare primary site of EGCT.⁴ Few cases have also been reported in the vulva, prostate, nasal cavities, bladder, uterus, paratesticular adnexa, kidney, and other sites.⁵⁻¹¹ The exact aetiology is not clear and they have a poor prognosis compared with their gonadal counterparts.

The aim of this report is to present primary extragonadal mixed germ cell tumour of the face in a Nigerian girl because of its rarity and diagnostic changes as well review available literature on the subject.

CASE REPORT

A 3-year old girl presented with her parents to a peripheral private hospital complaining of a progressively enlarging painless right facial swelling which started 6 months earlier. The swelling was initially small and slowly increasing in size but grew rapidly over the 2 months before presentation. On examination, the swelling measured 7x6x5cm over the right cheek and was ill-defined, firm in consistency, non-tender, non-fluctuant, and non-pulsatile. Other systemic physical examinations were unremarkable. Routine laboratory investigations were normal. Computed tomography (CT) scan and other radiological investigations were not done.

Incisional biopsy of the mass was done and the tissue was sent for histopathological assessment. The laboratory received irregular greyish white tissue measuring 3x2x1cm and the sample was reported as extragonadal mixed germ cell tumour with yolk sac tumour as the primary component and mature teratoma as the secondary component (figures 1, 2 and 3).

Based on the histopathology result, she was referred to our tertiary health institution for further management and follow up. The surgeons reviewed the patient and counseled the parents about the treatment modalities. She was given a two-week appointment

because of their financial challenges. However, she did not come back for review until 5 months later when she re-presented with complications. She was admitted and relevant investigations were requested. She died two days after commencement of treatments. The parents declined autopsy despite counseling.

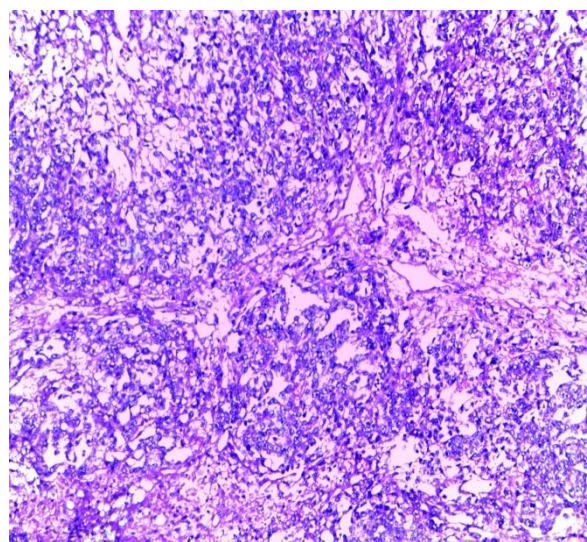


Figure 1: Yolk sac tumour shows Schiller-Duval bodies and small cystic spaces arranged in reticular pattern lined by cells with hyperchromatic nuclei and prominent nucleoli. (H&E X40; low magnification)

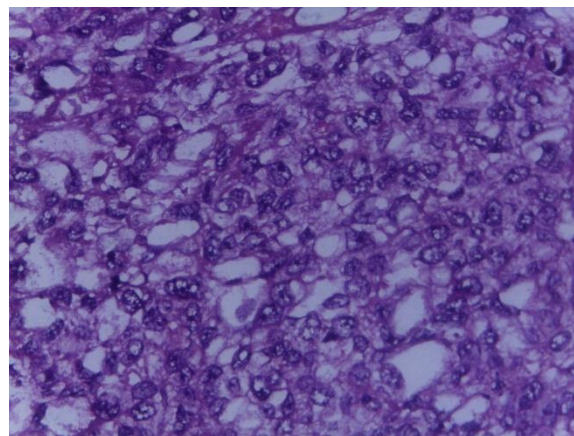


Figure 2: Yolk sac tumour shows Schiller-Duval bodies and small cystic spaces arranged in reticular pattern lined by cells with hyperchromatic nuclei and prominent nucleoli. (H&E X100; high magnification)

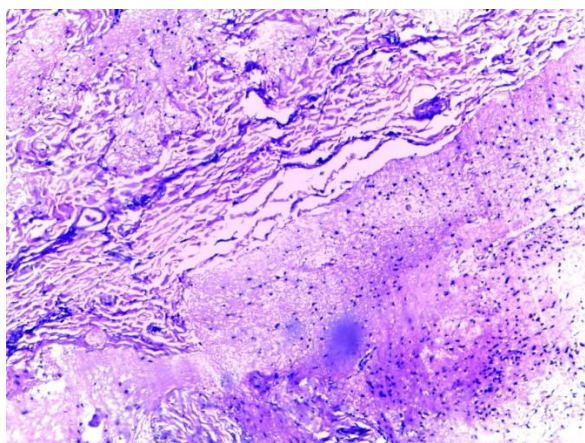


Figure 3: Mature teratoma shows meninges, choroid plexus, brain and adipose tissues. (H&E X40)

DISCUSSION

Germ cell tumours are called extragonadal when there is no evidence of a primary tumour in either the testes or ovaries. Primary EGCT comprise 1% to 5% of all germ cell tumours. The histogenesis and aetiology of these tumours are not clearly defined. EGCT more frequently affect young males but primary head and neck tumours usually occur in young females in keeping with the findings of the case reported.¹² Herein, we described a case of extragonadal mixed non-seminomatous germ cell tumour composed predominantly of Yolk sac tumour and a rare component of mature teratoma. The case is uncommon and this is the first case seen in our centre. The rarity of the case usually results in diagnostic challenges.

During the perinatal and childhood periods, teratoma is the most commonly diagnosed EGCT with equally documented number of mature and immature teratomas.¹³ Even though extragonadal Yolk sac tumours are uncommon and are particularly seen in sacrococcygeal, mediastinal, intracranial and retroperitoneal sites, they are still the commonest histological subtype among malignant EGCT, occurring mostly together with a teratoma and rarely alone. Head and neck region is an extremely rare site for primary extragonadal mixed germ cell tumour with very few documented cases.¹⁴ Currently 10 morphologic histological

appearances of yolk sac tumour are recognized by the World Health Organization namely microcystic, macrocystic, solid, glandular, papillary, endodermal sinus, myxomatous, polyvesicular, hepatoid and enteric pattern. This makes its diagnosis sometimes more difficult especially if the biopsy is small. Extragonadal germ cell tumours are extremely aggressive with high tendency for recurrence and metastasis especially to the lung. Spread to liver, bones and lymph nodes have also been documented.¹⁵

Tissue biopsy is essential for making a definitive diagnosis of EGCT. Some patients present with classical features of germ cell tumour while some present with an undifferentiated tumour with no distinctive germ cell features making the diagnosis difficult.

Surgical resection of the tumour mass was the standard treatment modality for EGCT in the past, but recently with the introduction of chemotherapy, the prognosis has dramatically improved like that of its gonadal counterpart. Prognosis of EGCT depends on the age of the patient, site of the tumour and the histological type. In adult patients with EGCT, histological type is the most important prognostic factor with seminomas having a long-time survival rate of about 90% while non-seminomatous tumours have about 45%.¹⁵

CONCLUSION

Extragonadal mixed germ cell tumour is an extremely rare neoplasm which is aggressive with high metastatic tendency. Therefore, timely diagnosis and early treatment is important when there is a high index of suspicion by the managing physician and the pathologist. Gonadal examination should be performed in all patients with extragonadal germ cell tumour for possible primary gonadal germ cell tumour.

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