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Rapidly Growing High-Grade Immature Ovarian Teratoma in A School-Age Child: Case Report of Successful Management of A Giant Prepubertal Mass.

Kasarachi Pauline Akowundu[1,3], Sampson Chukwuemeka Aliozor[1,4], Olubumi Ogein[1,5], Temitope Ebenezer Ogunsanya[2], Oluwaseye Oyeniran[1,5], Oluwa Tunde Odunafolabi[1,3], Olatubosun Olakunle[1,6], Lere Peter Oluwadare[1,3],Joshua Awofeso[1,7], Oyindamola Olabisi Aderinto[1,8], Israel Odutola Odetunde[1,3].

Department of Pediatrics[3], Surgery[4], Obstetrics and Gynaecology[5], Anesthesia[6], Radiology[7]and Family Medicine[8]
Redeemers Health Village (RHV), Redemption City, Mowe, Ogun State, Nigeria.[1]
Department of Histopathology, Federal Medical Centre, Ebute-Metta, Lagos State, Nigeria.[2]

Correspondence; **Kasarachi Pauline Akowundu**,
Redeemers Health Village, Mowe, Ogun State.

ABSTRACT
Ovarian teratoma is the most common germ cell neoplasm. It usually occurs in the first two decades of life. The immature teratoma comprises less than 1% of ovarian cancers and less than 1% of ovarian teratomas. Though immature ovarian teratoma can be aggressive, a high index of suspicion, early detection, and timely intervention with complete tumor excision are helpful to an eventual good outcome. Given the rarity, atypical age at presentation, and the grade of our case, it's being reported with a literature review.

KEYWORDS: *Mature, Immature; Ovarian, Teratoma, Tumor*

I. INTRODUCTION

Teratoma is derived from the Greek word "teraton, " meaning a monster. Immature Ovarian teratoma (IOT) is a specific histological subtype of germ cell tumor (GCT) which contains immature tissues derived from the 3 embryonic layers: the mesoderm, the endoderm, and the ectoderm.[1] IOTs contain anaplastic immature elements and most frequently neuroepithelial tissue.[2] Its grading depends on the amount of immature neural

elements, which is of prognostic value for the overall outcome. It is common in the younger age group, usually during the first two decades of life, with a peak incidence between 15 and 19 years.[3] Our patient is prepubertal with a huge, high-grade immature ovarian teratoma, which is rare. Given the rarity, atypical age at presentation, and the grade of our case, it's being reported with a literature review.

II. CASE PRESENTATION

A 10-year-old female who presented with a lower abdominal mass of about 1 month duration and was said to be progressively increasing in size. At the onset, there was no pain, but 2 weeks into the illness, she developed this dull pain, non-radiating, no known aggravating or relieving factor, not worse at any time. It was also associated with easy satiety and intermittent, non-bilious vomiting, not bloody, and post-prandial. There is no change in bowel habit, but weight loss is associated, as evidenced by the loosening of previously fitted clothing. There was no fever, drenching night sweat, no chronic cough, bone pain, and no mass in any other part of the body. There was no vaginal bleeding, she is yet to attain menarche. No urinary symptoms, family history of similar illness, nor family history of malignancy. On examination, she was not in any form of distress, not febrile, not pale, anicteric, not cyanosed, and had no pedal edema with stable vital signs. The abdomen was asymmetric with a huge intra-abdominal mass extending from the pelvis to the epigastrium, laterally extending to the lumbar regions, but more on the right. The mass was firm, dull, and mildly tender, showing some mobility on the horizontal axis but not on the longitudinal axis. It measures about 25cm across the lateral dimension. Hematological, renal function tests, and viral serology were within normal limits. Serum tumor markers were essentially within normal limits except CA 125. Abdominopelvic ultrasound showed a large cystic multiseptated right adnexa mass with mural nodules. The mass measures 223x131mm. The mass was indistinguishable from the right ovary, and it showed moderate vascularity on Doppler interrogation. The left ovary was sonologically normal, and there was minimal fluid in the pouch of Douglas.

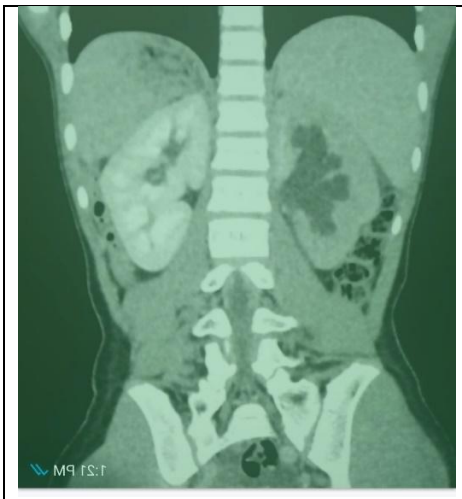


Figure 1

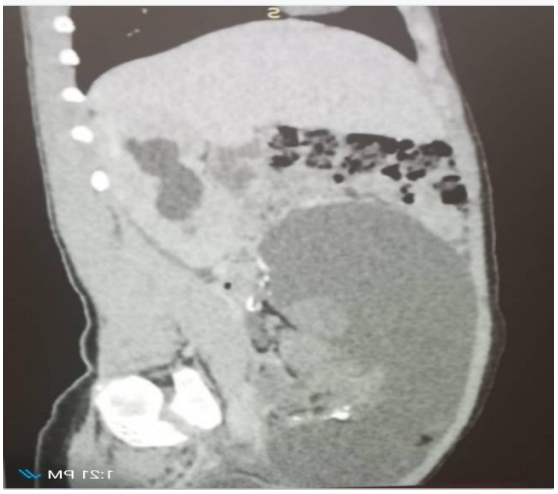


Figure 2

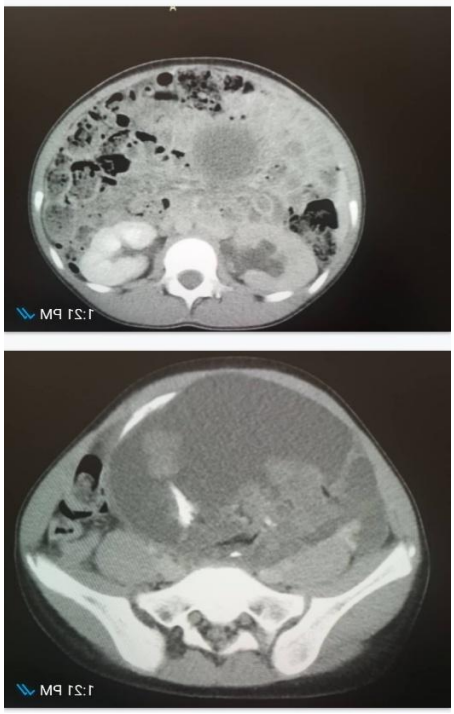


Figure 3A(above), 3B (below)

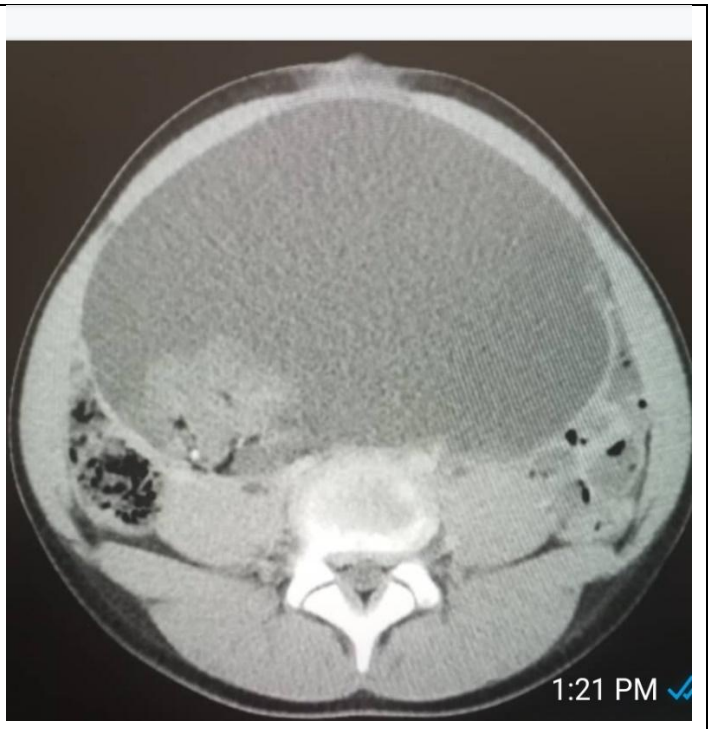
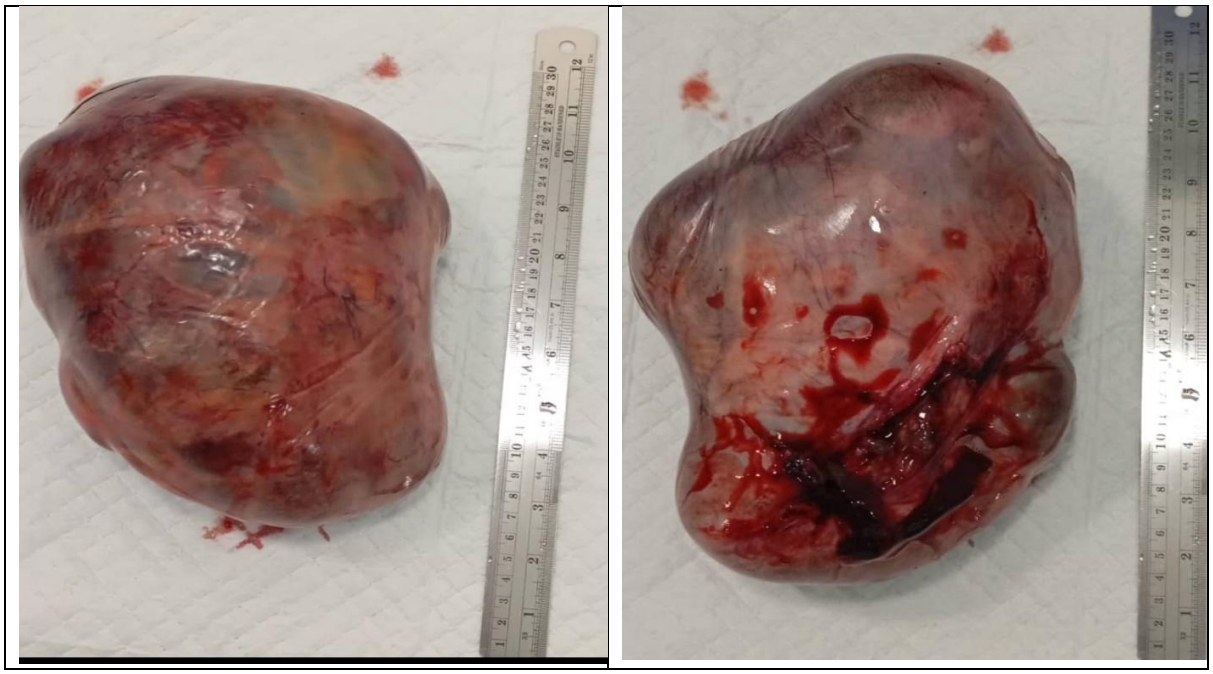


Figure 4

Abdomino-Pelvic CT Imaging

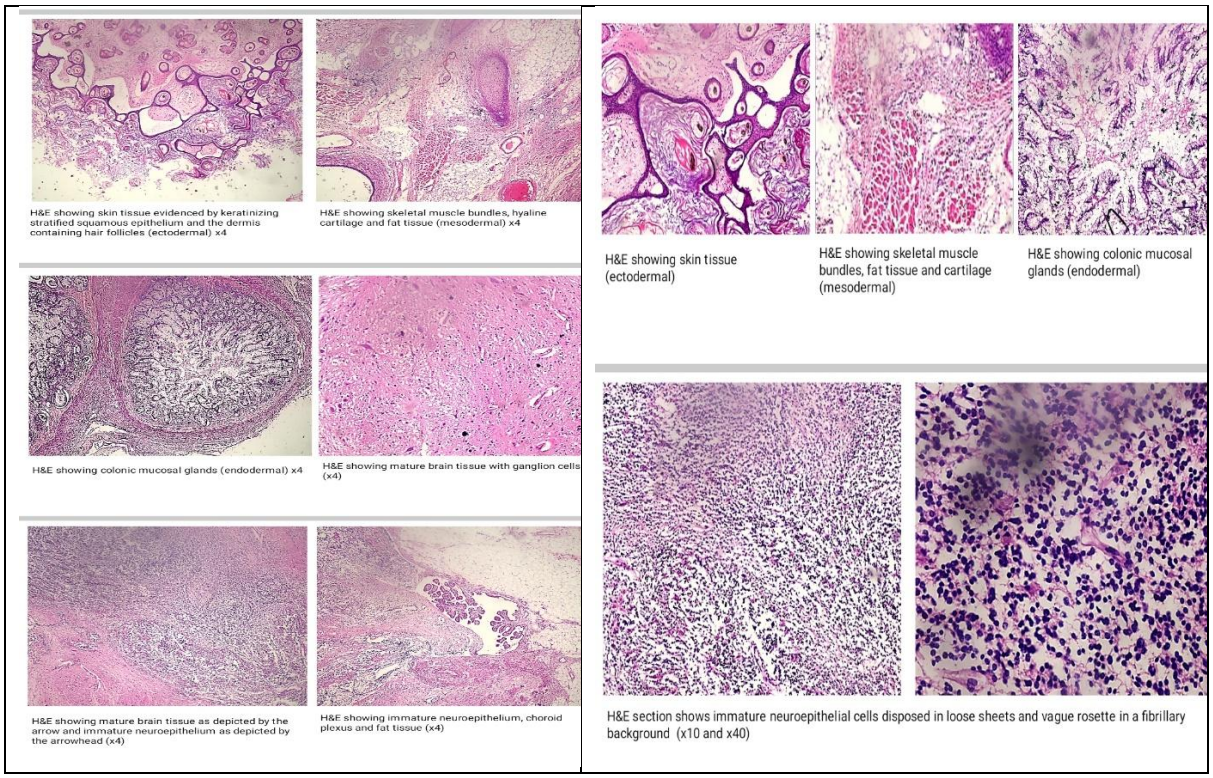
Figure 1-3A: demonstrates grade 2 hydronephrosis
Figures 2,3B, and 4 show a huge mass, predominantly cystic with some solid components
Figure 2-3A: shows a huge mass with marked bowel displacement superiorly.

The right kidney was enlarged, measuring 121 x 63mm, with mild dilatation of the pelvicalyceal system. The proximal ureter was dilatated, with no cortical atrophy, calculi, or mass lesion. The contralateral kidney was essentially unaffected. Other abdominal viscera were normal. Abdominopelvic CT scan correlated with the findings of ultrasonography as seen above. A complex abdominopelvic mass from the right adnexa measuring 223 x 100 x 160 mm was seen. The mass is predominantly cystic with some areas of discontinuous rim and septate calcifications, focal fat components, and several solid-enhancing nodules. The mass was compressing on the right urethra, resulting in grade 2 hydronephrosis. The left ureter is normal, and there was also moderate ascites. No adenopathy or areas of peritoneal fat stranding were noted. Other intra-abdominal viscera were within the normal limits. Intraoperative findings included an asymmetrically distended abdomen with a huge multilobulated cystic mass from the right ovary. Both tubes and the uterus were grossly tumor-free. There was no significant ascites, no nodal involvement, and a grossly uninvolved omentum unattached to the mass. There was a complete excision of the right ovary, sparing the ipsilateral fallopian tube that was macroscopically free. She had an uneventful postoperative period.



Picture of the huge mass

The histopathologic report showed an enlarged encapsulated cystic mass weighing 2500g and measuring 24 x 17 x 13cm. The surface was congested, and the cut section showed a huge cystic cavity containing greyish-brown secretion and focal solid areas in the inner lining with occasional hair strands. Microscopic examination revealed a malignant germ cell neoplasm composed of cells from the three germ layers with immature neuroepithelial cells in various proportions. There were skin tissues with adnexal structures, fat lobules, fetal hyaline cartilage, bone, choroid plexus, brain, colonic and respiratory tissues. The neuroepithelial component consists of embryonal cells disposed in sheets, tubules, and rosettes in >3/low power field. These confirmed the diagnosis of Immature teratoma Grade 3 (High Grade).



Histological slides of the lesion.

She was discharged on day 3 post-operation. The patient was counseled on the modalities of the next phase of care, and she is being closely followed up through active surveillance using clinical and laboratory investigations like Full blood count, urinalysis, liver and kidney function tests, tumor markers, and abdominopelvic ultrasound 3 monthly. She has been followed up for 6 months, and still ongoing, but no sign of recurrence.

III. DISCUSSION

Teratoma can be benign or malignant, and can also be classified as mono, bi-, or tri-dermal (ectoderm, mesoderm, or endoderm), depending on the number of germ layers present.1 Though teratomas occur commonly in children, IOT is relatively rare. [4] These tumors differ from Mature Ovarian Teratoma (MOT) is both histologically characterized by the presence of immature tissue and clinically by their more aggressive behavior.[2] The clinical manifestations of this disease are nonspecific. Generally, most patients with this tumor are asymptomatic until it reaches a considerable size, and this is also true for the child.[3] It tends to grow rapidly and may manifest as a pelvic or lower abdominal mass, which was the same location as our index case. As the tumor expands, it gives rise to pressure symptoms, abdominal heaviness, and dull pain, or it may undergo torsion, causing acute abdominal pain.[5] Our patient was already having pressure symptoms as evidenced by the easy satiety, vomiting, and abdominal pain with radiologic features of a grade 2 right hydronephrosis. Peritoneal implants and metastasis are not infrequently present at operation for the removal of the primary tumor of IOT, but our patient did not have any of such.[3,5] Additionally, these tumors are usually confined to the ovary; Immature teratomas have been found to invade their capsule, adhere to the surrounding structures, and later spread throughout the peritoneal cavity, initially to the retroperitoneal, para-aortic, and more distant lymph nodes and involve other organs, including the lungs and the liver.[3] However, our patient had neither adhesions nor distant metastasis, and the tumor capsule was intact. It has also been found that biopsies of macroscopically normal-appearing tissues were negative for tumor spread in all cases; hence, in our index case, grossly normal tissues were not biopsied.[6] Microscopic examination reveals tissues from all three germ layers, with most MOT containing elements that have undergone complete somatic differentiation, but IOT contains elements with only partial somatic differentiation, similar to embryonic or fetal tissue.[1] IOTs are graded based on the degree of cellular immaturity and the proportion of

immature neuroepithelium.[7] A higher proportion of immature neuroepithelium highly correlates with the tumor grade and prognosis.[7] Our patient had grade 3 IOT, which is a high grade. They macroscopically appear as a large solid mass with necrotic and hemorrhagic areas on cut sections, which was at variance with our patient, who had a huge cystic mass with focal solid areas.[8]The microscopic analysis shows immature neuroepithelium, which can be spindled(sarcomatous) or with rosette, pseudo-rosette, and primitive tubule formation.[5,9] In the case of our patient, they were displayed in sheets, tubules, and rosettes with greater than 3 per low-power field. Gonzalez-Crussi grading system is based on the amount of immature tissue within the tumor, having grade 0 as mature(benign), grade 1(<10% immature) probably benign, grade 2 (10-50% immature) possibly malignant, grade 3(> 50% immature), frankly malignant and this was observed in our patient.[10] Among the numerous tumor markers for germ cell tumors, the most commonly used are the CA 19-9 and Alpha-fetoprotein. These markers are of great value in screening, diagnosis, and treatment monitoring.11 They are within normal in our patient, both on presentation and on follow-up. However, CA 125 was 71.84 U/ml elevated above the <35.0 of the reference range. This value was reduced to normal during follow-up. The initial management of IOT is surgery, with further treatment dependent on the surgical findings.[12] For optimal outcomes, the management of IOT in pediatric patients should consist of a multidisciplinary team of pediatric surgeons, pediatricians, histopathologists, and gynecologists with a special interest in paediatric gynecology and gynaecologic oncology experts. The tumor size can be large at the time of diagnosis, like our patient, whose tumor weighed 2.5kg and measured 24cm in its widest dimension. Tumor size does not influence staging and does not affect prognosis.[2] Current treatment recommendations for IOT in children involves the minimal procedures required for staging, which include 1) removal of the primary tumor, without violation of the tumor capsule if intact at the time of surgery (noting that spontaneous, non-iatrogenic rupture may already have occurred; if so this should be carefully documented); 2) inspection and biopsy of any abnormal-appearing peritoneal surfaces, lymph nodes, omentum, and contralateral ovary; and 3) peritoneal fluid cytology. [13] In our patient, the mass was carefully excised en masse without violating the tumor capsule, and the peritoneal surfaces, omentum, and contralateral ovary were inspected and found to be grossly normal. Also, no significant ascites and lymph nodes. The tumor grade is an important risk factor for relapse across all age groups, while the stage is the most important factor for prognosis.[3,14] Patients with stage I tumors are often treated with surgical resection of the mass due to a favorable prognosis and very low risk of relapse, and do not require adjuvant treatment, however, postoperative review and close follow-up are recommended.[14]Postoperative chemotherapy is an indication for extra-ovarian disease.[14]Stage III, grade 3 patients are at ~50% risk of recurrence compared with ~10% risk for all other stage /group combinations. [14] Our patient had a stage I disease, which published evidence supports surgery alone with surveillance for all stage I patients, regardless of tumor grade. [15,16] This is what we offered our patient.

IV. CONCLUSION

Immature ovarian teratoma is a rare and complex neoplasm comprising <1% of ovarian cancers, occurring primarily in young females. The presentation can be subtle and can result in pressure symptoms. Diagnosis is necessary in grading, staging, and choosing a treatment modality. Surgery is the mainstay of therapy. In conclusion, management of ovarian teratoma and other germ cell tumors should be multidisciplinary. Though immature ovarian teratoma can be aggressive, a high index of suspicion, early detection, and timely intervention with complete tumor excision are helpful to an eventual good outcome

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