Case Report

A rare case of ovarian dysgerminoma in a 6-year old child in Lagos: A case report

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ABSTRACT

Dysgerminoma is the most common malignant germ cell tumor in children and adolescents. Most cases occur in the 2nd and 3rd decades of life, but 10% of cases occur in the 1st decade of life. The rarity of this malignancy in prepubertal children and the recognized controversies in its management prompted this case report. The case of a 6-year-old girl who had laparotomy and unilateral adnexectomy with subsequent histological diagnosis of an ovarian dysgerminoma was presented. Prognosis depends on the stage of the tumor at presentation, and conservative surgery should be the aim during the initial treatment. Adoption of multidisciplinary management within an oncology board with joint decision on the need for adjuvant chemotherapy for early stage diseases will confer good prognosis.

Key words: Germ cell tumor; malignant; management; multidisciplinary.

Introduction

Solid ovarian tumors are uncommon in the pediatric population, but when occurred, they are a major source of anxiety for the patients and their family. Pediatric ovarian neoplasms account for an estimated incidence of 2.6 cases per 100,000 girls per year.[1] Ovarian malignancy in children and adolescents is reported in 10%–20% of all ovarian masses or neoplasms and comprises approximately 1%–2% of all childhood malignancies.[2] Dysgerminoma originates from undifferentiated germ cells that are similar to primordial germ cells, and it is identical to testicular seminoma. Two-third of malignant ovarian tumors in children and adolescents are germ cell tumors (GCTs).[3] Most cases occur in the 2nd and 3rd decades of life, but 10% of cases occur in the 1st decade of life.[4] Most patients with dysgerminoma (75%) are diagnosed with early stage disease, and thus surgery alone is curative, and the prognosis is usually excellent,[5] but there is a significant dilemma with the management of advanced disease in children and adolescents. A rare case of ovarian dysgerminoma in a 6-year-old elementary school pupil who presented to the gynecological oncology clinic of our hospital is being reported. Information concerning the frequency and pattern of malignant ovarian tumors including dysgerminoma in children is scant in most Nigerian literatures. The rarity of this malignancy in prepubertal children in Nigeria and the recognized controversies in its management prompted us to report our experience.

Case Report

The patient was a 6-year-old girl who resided with her parents in Lagos, Southwest Nigeria. She was brought to the gynecological oncology clinic by her mother following a referral from a private hospital with a 3-week history of left-sided lower abdominal pain and progressive abdominal

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swelling. Pain was insidious in onset, intermittent, dull in nature, and not severe enough to affect sleep or her daily activities. It was aggravated by sitting up and relieved by taking analgesics. Abdominal swelling was initially the size of an orange but progressively increased to reach the level of her navel. No history of chronic cough, but there was associated weight loss as evidenced by her clothes being oversized. There was no similar history in her siblings or parents. There was a history of vomiting of noncopious recently ingested meals on few occasions. The mother said her pregnancy was uneventful with no febrile illness or use of any unprescribed or illicit drugs during pregnancy. The child was delivered through spontaneous vaginal delivery at term at a private hospital. Pregnancy, labor, and puerperium were uneventful. Developmental milestones were attained at the appropriate times. An abdominopelvic computed tomography scan done on presentation at the referring hospital a week earlier was suggestive of a huge heterogeneous mass arising from the pelvis and extending superiorly into the abdomen. The mass was mainly solid with multiple areas within it. It had no calcifications within it and measured 12.5 cm × 8.10 cm × 7.5 cm [Figure 1]. Examination findings on presentation to us were that of a full abdomen with obvious suprapubic fullness and a 20-week sized abdominopelvic mass which was firm, non-tender, smooth, and mobile. Tumor markers including beta-human chorionic gonadotrophin and alpha-fetoprotein were essentially normal, but there was a 10-fold elevation in the level of lactate dehydrogenase (LDH). The findings and options of management were explained to the parents, and the child was planned for laparotomy and unilateral left adnexectomy (salpingo-oophorectomy) which she had 5 days later. Intraoperative findings [Figure 2] showed no ascitic fluid; a large left ovarian solid mass (15 cm × 12 cm × 10 cm) with grossly normal left tube; the right ovary and tube were also grossly normal; and minimal adhesion involving the left ovarian mass and omentum, otherwise the omentum was grossly normal. Her postoperative period was uneventful, and she was discharged home on the 4th postoperative day. The histological examination of the ovarian mass revealed a dysgerminoma with focal areas of necrosis without infiltration of the attached omentum. The child had no additional postoperative treatment and is currently being closely monitored with regular follow-up visits, physical examinations, ultrasound scan, and LDH assays. She has remained tumor-free for the past 3 years after her surgery.

Discussion

This report revealed the management outline of a 6-year-old Nigerian child with an ovarian dysgerminoma. Malignant neoplasms of the ovary in childhood are extremely rare and far less common than the benign ovarian tumors.[6] Ajani *et al*.[7] reported the rarity of childhood ovarian malignancies as only 24 cases of ovarian neoplasms were seen between the age group 0 and 14 years (<15 years of age) over a 22-year period (1991–2003) at University College Hospital, Ibadan. Dysgerminoma was found to be the most common primary malignant childhood ovarian tumor in Nigerian children,[7] and there was only one reported case in the Nigerian literature over the past 10 years (2006–2015).[8] These explain why there are only scattered reports on childhood ovarian malignancies in the literature and why there is a lack of specific pediatric treatment.

Ovarian lesions including malignancies can present with variable signs and symptoms, and most often, the clinical presentation does not differentiate benign tumor from a malignant tumor as found in the index case. Abdominal pain

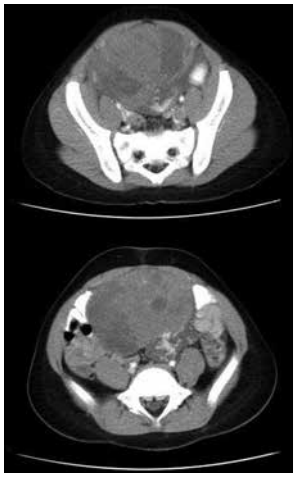

Figure 1: Abdominopelvic computed tomography scan showing the huge complex adnexal mass in the 6-year-old child

Figure 2: Intraoperative finding of ovarian dysgerminoma in the 6-year-old girl

was the most common symptom encountered.[8] A mobile, palpable abdominal mass was the most frequent physical finding,[8] as we also observed in our patient. Elevated LDH can be an early clinical sign of an ovarian dysgerminoma. However, it is not all dysgerminomas that produce LDH, and this is often regarded as a nonspecific finding. Our patient had a 10-fold increase in her LDH levels.

The management of childhood ovarian tumor must be balanced with the desire to maintain the child's reproductive and developmental potential. A definitive histologic diagnosis and a thorough understanding of the behavior of each of these tumors are necessary for appropriate therapy. Surgery is the initial step to obtain definitive diagnosis and provide initial treatment of patients with malignant ovarian GCTs.[9] Initial conservative management is, however, essential during this surgical treatment of pediatric ovarian tumors. A diagnosis from a frozen tissue section should not be used as the basis for radical surgery for a unilateral tumor in an otherwise unremarkable pelvis. If the lesion is a stage IA, just like in the index case, no postoperative therapy is indicated. However, if the dysgerminoma involves both ovaries, bilateral salpingo-oophorectomy and a full staging operation should be carried out; however, unless the uterus is involved by cancer, it should not be removed because *in vitro* fertilization with a donor ovum can be an option for the child in the future.[9] Metastatic disease demands complete cytoreductive surgery.

In the past, many stage I patients and all patients of higher stage were treated with radiotherapy because dysgerminomas have an inherently high degree of radiosensitivity. However, pelvic radiotherapy is associated with a high incidence of sterility, but with the development of effective chemotherapy regimens, radiotherapy is rarely indicated except for unusual metastatic patterns which is rarely seen in children. The current recommendation for adjuvant chemotherapy in the treatment of ovarian dysgerminoma is BEP (bleomycin, etoposide, and cisplatin).[10] This regimen has shown a sustained remission rate of 75%[9,10] and is currently the standard of care for the more advanced ovarian dysgerminoma in most centers including ours.[9]

The malignancy of pure dysgerminomas is relatively low grade, with survival rates of 80%–96%. The incidence of recurrence is, however, reportedly high, ranging between 33% and 50% in some studies with most recurrences seen within the first 1 year after therapy with extreme rarity after 2 years.[10] Surveillance visits following active treatment should, therefore, consist of history, physical and pelvic examination, imaging, and relevant tumor markers quarterly for the first 2 years and biennially for 3 additional years. The index patient was also having postsurgery follow-up surveillance which will last for the next 5 years. There are still some disagreements regarding the optimal treatment of patients with dysgerminomas who received conservative surgical therapy in childhood and who later completed their desired childbearing. Exploratory laparotomy with a hysterectomy and salpingo-oophorectomy has been suggested for these patients. Because of the theoretical potential for late recurrences, the rarity of these tumors and the unknown benefit of this surgical intervention, however, most clinicians do not recommend this method of treatment.

Conclusion

Ovarian dysgerminomas are extremely uncommon in children. Pain and abdominal mass are the most common modes of presentation. Prognosis depends on the stage of the tumor at presentation, and conservative surgery should be the aim during the initial treatment in children and adolescents. Adoption of multidisciplinary management within an oncology board with joint decision on the need for adjuvant chemotherapy for early stage diseases will confer good prognosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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