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Ovarian dysgerminoma in an adolescent: a case report

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Abstract

Introduction: Ovarian cancer is the second most frequent gynaecological cancer in Nigeria ranking next after carcinoma of the cervix. It has the highest case-fatality rate worldwide because of insidious onset, lack of effective screening methods and late presentation. This case of a sixteen-year old girl with a three-week history of abdominal pain which was later accompanied by abdominal swelling is a classic example of how dysgerminomas present.

*Method:*The presumptive diagnosis of an ovarian tumour was made after physical examination and this was later confirmed by ultrasound scan. Urgent laparotomy was carried out based on a suspicion of torsion of the pedicle of the cyst.

Result: At laparotomy, torsion of the pedicle with an intact capsule and imminent gangrene were found. The histology report revealed a malignant germ cell neoplasm (Dysgerminoma) with focal areas of necrosis without infiltration of the attached omentum.

Conclusion: She had conservative surgery (left oophorectomy) done. She, however, defaulted from further follow-up.

Keywords: *Ovarian, cyst, cancer, laparotomy, dysgerminoma.*

Résumé

Introduction: Le cancer de l'ovaire est le second cancer gynécologique le plus fréquent au Nigeria, après le cancer du col. Son taux de létalité est plus élevé dans le monde entier en raison de l'apparition insidieuse, de l'absence de méthodes de dépistage efficaces et de leur tardive introduction. Le cas d'une jeune fille de seize ans avec une histoire de trois semaines de douleurs abdominales qui, a plus tard résulté e en un gonflement abdominal est un exemple classique de la façon dont se présentent les Dysgerminomes.

Méthodes: Le diagnostic présomptif d'une tumeur de l'ovaire a été fait après un examen physique et a été confirmé plus tard par l'échographie. Laparotomie d'urgence a été réalisée sur la base d'une suspicion de torsion du pédicule du kyste.

Résultat: Lors de la laparotomie, la torsion du pédicule avec une capsule intacte et la gangrène imminente ont été trouvés. Le rapport histologique a révélé une tumeur germinale maligne (Dysgerminome) avec des zones focales de nécrose sans infiltration de l'épiploon ci-joint. Elle a subi une chirurgie conservatrice (à l'ovariectomie gauche). Mais, elle a, cependant, manqué de suivi par la suite.

Conclusion: Une fois diagnostiqué, le Dysgerminome répond bien au traitement, ce qui devrait être engagée dès que possible.

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Introduction

The cause of ovarian cancers is unknown but the commonest risk factor is a positive family history of ovarian or breast cancer [1]. Primary tumors of the ovaries may arise from the epithelium (82%), sex cord and germ cell (20%) areas. About 3 to 5per cent of all germ cell tumors (GCTs) are malignant with the most common being the dysgerminoma which also accounts for 4-5% of all ovarian cancers1. Ovarian dysgerminoma is a relatively rare tumour of the ovary, however, it is a malignant and aggressive tumour [2]. It is commoner in young women and adolescents [3] and it is known to be radiosensitive [4]. Adjuvant chemotherapy is sometimes used with the hope of preserving fertility in young women whenever the primary surgery is thought to be inadequate treatment considering the clinical staging of the disease. Simple oophorectomy can be performed with good results when the tumour is detected early. However, the tumour could be bilateral in some cases when some form of conservative surgery (excision biopsy of the second ovary) may be necessary. Many of these young patients usually present as a result of accidents to the cyst and/or abdominal mass.

We present this case of dysgerminoma in a 16-year old school girl who had laparotomy and fertility-preserving surgery but travelled back to her home town and was lost to follow-up.

Case report

A sixteen-year old , para 0+0 , secondary school student presented with right iliac fossa pain and abdominal distension of three and one week duration respectively. The pain was intermittent, non-colicky, non-radiating without any known aggravating factor. There was no associated history of fever, bowel symptoms, weight loss or vaginal bleeding. The only

treatment she had prior to presentation was oral analgesics. On examination, she was found to be ill-looking but she had no pallor, jaundice or pyrexia. There was a 24-week size abdomino-pelvic mass which was tender, firm and slightly mobile. Vaginal examination was not performed because of her virginal state. A presumptive diagnosis of torsion of an ovarian cyst was made and she had full blood count, electrolytes and urea and were all within normal limits. Abdominal ultrasound scan revealed a solid left adnexal mass, 126mm in diameter extending to the right side with an echogenic centre. The kidneys, ureters and bladder were normal and there were no detectable para-aortic nodes. She was admitted for exploratory laparotomy.

Laparotomy findings included scanty serous ascitic fluid, the omentum was attached to the left ovary which was about 18cm in its widest diameter. It was darkish, lobulated and highly vascular with its pedicle twisted and the capsule was intact. The left and right fallopian tubes and right ovary appeared normal. This was classified as International Federation of Gynaecology and Obstetrics (FIGO) stage 1A disease. She had left oophorectomy and partial omentectomy.

Patient made good clinical recovery from surgery and was discharged on the 7th post-operative day.

At histological examination, the morphology of the specimen revealed a bean-shaped, grayish-brown, encapsulated tumour weighing about 950g and measuring 14x11x10cm. The cut section showed an aggregate of moderate-sized uniform cells with large vesicular nucleus, prominent nucleoli and scanty to moderate cytoplasm. The cells were arranged in an organoid pattern as well as in nests or single files with a stroma of loose fibroblastic or dense fibrocollagenous cells and a conclusion of ovarian dysgerminoma. The omental biopsy was a piece of fibro-fatty tissue weighing 50gm and measuring 12x8x2cm. It showed intense infiltration by acute and chronic inflammatory cells with foci of granulomas with foreign body giant cells consistent with chronic granulomatous inflammation.

She was counseled along with her parents on the implications of the histology report and the need for adjuvant chemotherapy and out-patient follow-up but she defaulted from follow-up.

Ovarian dysgerminoma in an adolescent

Legend to the attached picture (Figure 1)

This shows an aggregate of moderate-sized uniform cells with large vesicular nucleus, prominent nucleoli and scanty to moderate cytoplasm. The cells are arranged in an organoid pattern as well as in nests or single files with a stroma of loose fibroblastic or dense fibrocollagenous cells. Haematoxin and Eosin staining. Magnification: ×40.

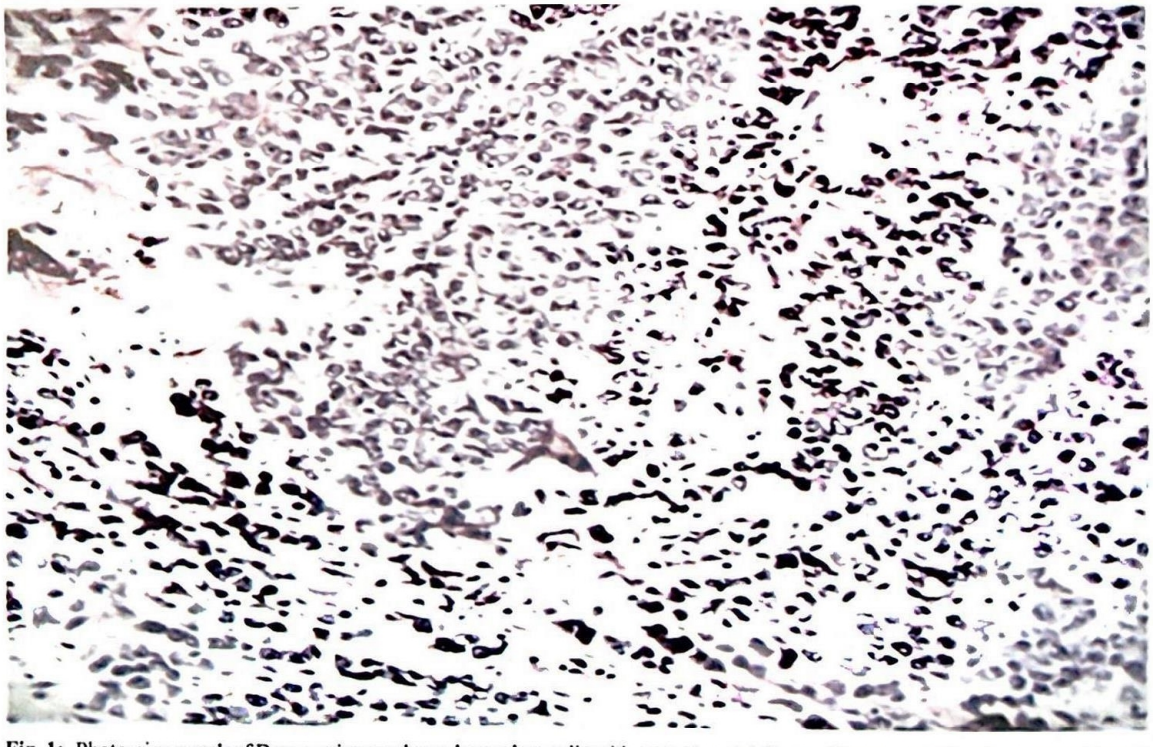

Fig. 1: Photomicrograph of Dysgerminoma shows large clear cells with central nuclei disposed in nests and insular pattern. There is lymphocytic infiltration of the intervening fibrous stroma

Discussion

Of the ovarian lesions, 97per cent are benign proliferations such as mature teratomas, the remaining 3 per cent are malignant [3]. Dysgerminomas accounts for 4 to 5 percent of all cancers of the ovary, but less than 1per cent of ovarian tumors overall. They are the most common malignant germ cell tumor occurring in the ovary, and these lesions are found most commonly in adolescents and young adults; in fact, approximately 60per cent of cases are diagnosed in patients younger than twenty years [4]. Unlike other germ cell tumors, dysgerminomas often occur bilaterally in approximately 10 to 20 per cent of cases.

Common signs and symptoms of ovarian dysgerminomas include abdominal/pelvic pain as in our patient, abdominal mass, fever, vaginal bleeding and, occasionally, ascites in that order. This patient had been experiencing the right iliac fossa pain three weeks prior to presentation. It is possible that she may have had some abdominal pain earlier which she attributed to indigestion as it is commonly thought in this environment. It was when the pain became unbearable (due to the ongoing torsion of its pedicle and the attendant ischaemia), that she reported to the school clinic. Her not noticing the swelling of the mass until a week prior to presentation is also typical of ovarian tumours due to the insidious growth of the mass. It would seem that it was the abdominal pain that alerted the patient to the presence of a swelling.

Extra ovarian tumor spread of dysgerminomas often involves the retroperitoneal and pelvic lymph nodes. In addition, haematogenous spread may occur; common sites of involvement are the lungs, liver, and bone [4]. Neither of this was found in this patient. A FIGO clinical stage of 1A was apportioned in this patient's case because of the unruptured capsule. There was no significant ascites even though peritoneal washings could have been sent for cytology to exclude of presence of malignant cells.

Dysgerminomas are known to be associated with elevated serum levels of lactate dehydrogenase (LDH). Although these tumors are thought to be hormonally inert, at least a case of precocious puberty with elaboration of high levels of beta-human chorionic gonadotropin, alpha-fetoprotein and oestradiol occurring in association with dysgerminoma has been reported [5]. In addition, elevated serum levels of neuron-specific enolase, calcium, inhibin, placental alkaline phosphatase (PLAP), and prolactin have been reported [6, 7, 8, 9]. These serologic elevations readily resolve following surgical excision; after the elevations resolve, the serum levels may be used as tumor markers to monitor for recurrence.

The prognosis and treatment of dysgerminomas depend on their pathologic and clinical stage. Patients with stage 1A disease (disease that is limited to one ovary) may be treated by unilateral oophorectomy alone. Although the patient defaulted, the surgical procedure performed and the non-invasion of the tumour capsule nor attached omentum may possibly suggest that the treatment was adequate in this case. The relapse rate ranges from 10 to 20per cent; the overall survival rate is 90 to100 per cent [10]. Patients who suffer relapses may undergo chemotherapy; the survival rate for such patients is greater than 90 per cent [9]. The 5-year survival rate is 96% if the tumor is confined to the ovary and 63% if extension occurs beyond the ovaries.

Radiotherapy and 3 to 4 cycles of adjuvant chemotherapy with cisplatinum, etoposide/vinblastine, and bleomycin are often reserved for patients with at least stage 1B disease (disease that is limited to both ovaries) or for those patients who suffer recurrences. For these patients, the results of therapy are similar to those of patients with stage 1A disease [11].

Despite the fact that most of the patients in our environment usually present in the late stages of ovarian cancer [12]; this patient presented early and was treated promptly. In conclusion, once diagnosed, dysgerminomas respond well to therapy and this should be instituted as soon as possible. The advantage is to spare patients from infertility and early mortality.

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