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Burkitts lymphoma of the ovary in Nigerian adults — a 27-year review

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Summary

Ten cases of adult Burkitts lymphoma managed over a 27-year period at the University College Hospital, Ibadan were reviewed. The incidence of adult ovarian Burkitts over the period was 1.3%. Menstrual abnormalities and abdominal masses were the most common symptoms while bilateral multinodular ovarian masses were the most constant findings at laparotomy. Diagnosis was mainly by histopathology. Surgery and chemotherapy were the main modalities of treatment. Mortality was high with cerebral metastases being the most common cause of death.

Résumé

Dix cas de lymphome de Burkitt chez l'adulte qu'on a traité pendant le période de 27 ans à l'University College Hospital, Ibadan ont été réexaminé. La fréquence de lymphome de Burkitt affectant l'ovaire pendant cette période était 1.3%. Les anomalies menstrueles et l'anomalie de tumeurs abdominales font les plus fréquents symptômes tandis que les tumeurs d'ovaire bilatérales et multinodulaires font les plus constants résultats à la laparatomie. Le diagnostic était fait par l'histopathologie. La chirurgie et la chimiothérapie font les modalités de traitement. La mortalité était élevé et les métastases cerebrales étaient le cause plus commune de décès.

Introduction

Malignant ovarian tumours constitute 6.1% of all female cancers in Ibadan [1]. In children and adolescents (0–20 years) in Ibadan the incidence is 13.6% [2]. It is much higher than the 6% quoted by Breen and Maxson [3]. This is said to be due largely to the preponderance of ovarian Burkitts lymphoma which was found to account for over 55% of ovarian neoplasms in Nigerian children [2]. Most of the ovarian Burkitts lymphomas occur between the ages of 5 and 14 years with few occurring after the age of 14 [2,4]. Williams *et al.* [5] stated that the sporadic cases of Burkitts tumour reported in the U.S.A. ranged from 2.3 to 27.0 years of age with a mean age of diagnosis of 10.2. This mean however, is said to be 2 years more than that reported from regions of Africa where the tumour is endemic.

Burkitts lymphoma is the most common childhood malignancy in the black African population of most of the tropics [6], and frequently the ovary is involved [4,7]. Adult Burkitts lymphoma is rare and adult ovarian Burkitts rarer still. Nelson *et al.* [8] reported on six cases of malignant lymphoma involving the ovaries over a 20-year period in patients aged between 19 and 58 years and found four more cases in the literature in the same age range. However, these cases were non-Burkitts lymphomas. Armon [9] reported on two cases associated with pregnancy while Junaid [2] reported on seven cases in patients aged between 15 and 19 years over a 20-year period.

The literature on adult ovarian Burkitts is scarce and experience with this condition is therefore limited. The objective of this report was to determine the prevalence of adult ovarian Burkitts among women with ovarian tumours at the University College Hospital (UCH), Ibadan and to review the clinical presentation and operative findings of all the cases seen.

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Subjects and methods

The cancer register in Ibadan between 1960 and 1987 was reviewed and all cases with histological diagnosis of malignant ovarian tumours were identified. Case records of those with a histological diagnosis of Burkitts lymphoma and who were aged 18 years and above were retrieved from the medical records for analysis. The clinical presentation, physical findings, treatment modalities and complications were analysed.

Results

During the study period, there were 10 cases of adult ovarian Burkitts among 759 malignant ovarian tumours managed at the UCH, thus giving an incidence of 1.3%. Five of these were between the ages of 18 and 25 years, 3 were between 26 and 35 years, one was 55 years while the other was 60 years old. Four were nulliparous, two of parity 1–2 and three of parity 3–4. One was para 6.

Two of the ten cases were seen at the surgical out-patient department while four were seen in the gynaecology out-patient clinic. Two presented as cases of acute abdomen to the accident and emergency department, while two others were referred to the gynaecology clinic from the general and medical out-patients.

The most common symptom at presentation was menstrual abnormality, occurring in seven cases. One had had irregular menses for 3 years, then became amenorrhoeic 6 months before presentation. Another had had irregular periods for 6 months and amenorrhoea for 4 months. Other symptoms are shown in Table 1. Abdominal pain was the symptom in six patients while five presented with abdominal swelling and four with weight loss. Infertility was one of the presenting complaints in two patients while three presented with recurrent fever. Vomiting and jaw swelling were present in one case each.

Amongst the physical signs, multiple abdominal masses were the most common, being present in seven patients. Other findings included ascites (3 cases), significant peripheral lymphadenopathy (2) and adnexal masses (4). Low grade pyrexia and pallor were present in two cases each.

Table 2 shows the findings at laparotomy.

Table 1. Presenting symptoms among 10 cases of adult ovarian Burkitts lymphoma seen at the University College Hospital, Ibadan, from 1st January 1960 to December 31st 1987

Symptom	Number of cases
Menstrual patterns	
Irregular periods	5
Secondary amenorrhoea	3
Menopause	2
Abdominal pains	6
Abdominal swelling	5
Weight loss	4
Fever	3
Infertility	2
Vomiting	1
Swelling of the jaw	1
Others*	3

*Backache, breast lumps and weakness.

Table 2. Surgical findings in 10 cases of adult ovarian Burkitts seen at the University College Hospital, Ibadan in a 27-year period

Finding	Number of cases
Bilateral ovarian masses	9
Unilateral ovarian mass	1
Ascites	4
Ileocaecal masses	3
Nodal enlargement	3
Splenomegaly	1

Bilateral multinodular ovarian masses were found in nine cases while only one had a unilateral multinodular ovarian tumour. The masses were friable, heavy and on long pedicles. One of the patients who presented to the accident and emergency department had torsion of one of the ovarian masses. Ileocaecal tumours (causing intestinal obstruction in two cases and intussusception in one case) were identified in three cases while three patients had enlarged paraortic and pelvic lymph nodes. Ascites occurred in four cases and one of these was haemorrhagic while the others were straw-coloured. The spleen was enlarged in one case.

The treatment offered to the patients varied considerably and depended on the surgeon. Total abdominal hysterectomy and bilateral

salpingo-oöphorectomy was the most common form of surgery performed. It was performed in six cases, four of which also had total or partial omentectomy. Resection of ileocaecal tumour and intestinal anastomosis was also offered to three of these cases. Of the others, an oöphorectomy and cystectomy were offered to one, bilateral wedge resection to another while left oöphorectomy and biopsy of the right ovary was the surgical treatment in one patient. The gynaecologists performed hysterectomy on their cases while the general surgeons did resections and sampling of nodes.

Of the ten cases, one died shortly after surgery from severe anaemia. The other nine were given cyclophosphamide after histological diagnosis of Burkitts lymphoma. Figure 1 shows the typical histological features as seen in the most recently managed patient. Two died within 1 year of surgery, the same number died within 2–3 years post-surgery and three were followed for up to 5 years. One case failed to report to the clinic after hospital discharge but the one recently managed is being followed-up.

Cerebral involvement was the most common cause of death. Of the five deaths, cerebral involvement accounted for three although one also had severe anaemia. Sepsis was the immediate cause of death in one case and anaemia alone in another. All had a post-mortem examination.

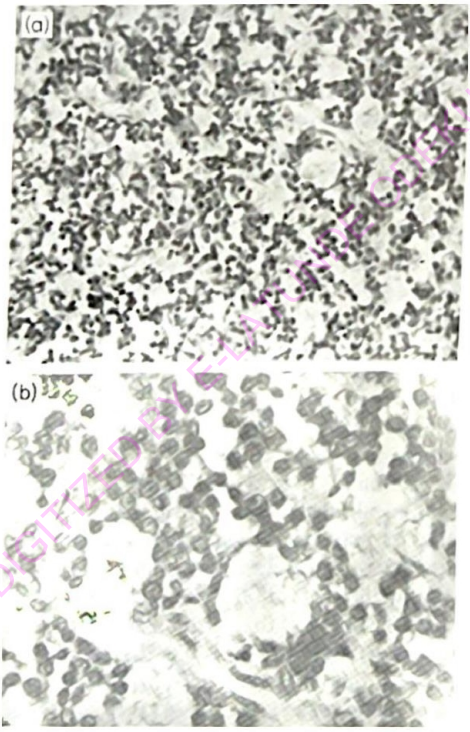

Fig. 1. Section of ovarian Burkitts showing the typical starry-sky appearance (a) ×100; (b) ×220.

Discussion

Burkitts lymphoma is one of the most common childhood tumours in Ibadan and the tropical region [2,10]. It is able to manifest itself in many organs with the face and the abdomen as the commonest sites. In the abdomen, the ovary is one of the most commonly afflicted organs [10]. Although ovarian tumours are rare in childhood, ovarian Burkitts constitutes 7.0 of the 13.6% of childhood ovarian neoplasms in Ibadan [2]. Ovarian Burkitts is, however, a very rare tumour in the adult with few reported cases in the literature [2,8,9]. This is probably due to the fact that by adulthood, most of the patients who would have been susceptible would have been eliminated [11].

The rarity of this tumour in the adult genital tract is clearly exemplified by the common misdiagnosis in the few reported cases [4,12]. Junaid [2] found that most of the cases he reviewed were diagnosed at autopsy. Ninety per cent of the cases in this series were diagnosed from histology. Those that occurred in young adults were commonly misdiagnosed as dermoid cysts. Suspicion of Burkitts lymphoma was only suggested in one case because she had typical radiological characteristics of jaw Burkitts.

The symptomatology of ovarian Burkitts, as illustrated in this study, is varied. Menstrual abnormalities appear to be the most common symptom. Infertility which is closely related to ovarian function was a form of presentation in two patients and could be due to the rapid replacement or the ovarian tissue by lymphoid cells. In most of the tumours removed, virtually no normal ovarian tissue was identified. Such patients will therefore present with hypomenorrhoea, amenorrhoea and infertility. Pregnancies have, however, been reported in some patients with ovarian Burkitts [9]. It is possible

that some of the patients might have been pregnant after the tumour had developed. In such cases, it is either that only one ovary was afflicted or replacement of normal ovarian tissue was yet to be complete.

Though Burkitts lymphoma of the adult ovary is rare, this review has revealed certain laparotomy findings that appear almost unique. Unlike most advanced malignant tumours ascites was present in only 40% of cases. However, this was bloody in only one case. The tumours were bilateral in 90% of cases and multinodular in all the cases. Of significance is the fact that all were non-cystic and felt firm in consistency. They were quite vascular, had long pedicles and little or no identifiable normal ovarian tissue. This latter finding is at variance with that in other ovarian tumours. The presence of such features at laparotomy in an adult from an area with a high prevalence of Burkitts should draw a suspicion of ovarian Burkitts. The rarity of abdominal and pelvic lymphadenopathy in this study supports the view that this disease is extranodal in origin [2]. Of the other tumours affecting the ovaries, ileocaecal invasion is very rare, however, this was a common finding in this series. The presence of multinodular ovarian and ileocaecal tumours is thus highly diagnostic of Burkitts lymphoma. Tuberculosis of the abdomen and primary ileocaecal tumours with ovarian secondaries must, however, be excluded. As response to chemotherapy is good, as shown in this series and others [13], a high index of suspicion based on the clinical and surgical findings is required. This must, however, be supported by skilled histopathology for confirmation.

The treatment often advocated for malignant ovarian tumours is surgery and chemotherapy. In this series, the treatment most commonly offered was total abdominal hysterectomy, bilateral salpingo-oöphorectomy and omentectomy followed by chemotherapy with cyclophosphamide. The chemotherapy was offered only after a histological diagnosis was made. Carbone *et al.* [13] recommended surgical reduction of tumour bulk in patients with extensive disease when possible, followed by cyclophosphamide (40 mg/kg) intravenously every 2 weeks for six doses. Response to this regime in this series was shown to be fairly satisfactory. This compares favourably with results of Carbone *et al.* [13] from treatment of 12 patients (mainly children) with this regime. In the two patients who presented with recurrences within 6–9 months, after the initial treatment, a repeat course of cyclophosphamide yielded satisfactory remision. It is therefore believed that cyclophosphamide can be used for primary treatment and for recurrences. Subsequent recurrences should, however, be treated by other drugs as recommended by Burchenal [14] as increasing resistance may make response unsatisfactory.

Since this tumour is rapidly growing, and highly malignant, it would seem reasonable to offer young patients radical surgery as complete replacement of normal ovarian tissue occurs frequently. However, chemotherapy induces rapid tumour regression, and following this resurgence of normal ovarian tissues may occur with maintenance of reproductive function. Based on these premises, we believe that there is a place for conservative treatment in young patients, but this will depend on the results from a close follow-up of those who had wedge resection or unilateral oöphorectomy by hormone assays, ultrasound scans, nuclear magnetic resonance and second-look laparotomies and ovarian biopsy over long periods.

Ovarian Burkitts lymphoma has remained controversial as regards its origin. While Haplin [4] described one case which he considered to be primary ovarian Burkitts, Nelson *et al.* [8] and Serov *et al.* [15] questioned the basis for such conclusions by pointing out that lymphocytic aggregates were never found in the normal ovary. Their conclusion was that all ovarian Burkitts were secondary. Woodruff *et al.* [16] however, reported that lympoid tissue may occasionally be present in the hilus or medulla of the ovary and it is possible, although rare, that the ovary is the primary site of a lymphoma and also Burkitts lymphoma has a predominantly extranodal origin and a wide anatomical distribution. Although from this series it may be concluded that some of the tumours were primarily ovarian (as there was no evidence of other organ involvement) it must be cautioned that such an assumption may be misleading. It is possible that microscopic Burkitts tumours from other distant sites might have metastasized to the ovary, and unless highly sensitive and very specific diagnostic techniques are available to screen for such microscopic primary sites, the controversy will

remain. The suggestion of Zeigerman *et al.* [17] that such controversies be disregarded for now may be acceptable but should not limit the quest for such a distinction. It is probable that differentiation between primary and secondary tumours may shed more light on aetiology, response to therapy and prognosis of this common childhood, but very rare adult, ovarian tumour.

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References

1. Abioye AA. The Ibadan Cancer Register 1960–1980. In: Olatunbosun OA, ed. Cancer in Africa, Proceedings of a Workshop of the West African College of Physicians. Ibadan: Ibadan University Press, 1981:1–32.
2. Junaid TA. Ovarian neoplasms in children and adolescents in Ibadan, Nigeria. Cancer 1981;47: 610–14.
3. Breen JL, Maxson WS. Ovarian tumours in children and adolescents. Clin Obstet Gynaecol 1977;20:607–23.
4. Haplin TF. Gynaecologic implications of Burkitts tumour. Obstet Gynaecol Survey 1975;30: 351–8.
5. Williams WJ, Butler E, Erslev AJ, Rundles RW. Burkitts lymphoma. In: Haematology. New York: McGraw-Hill, 1970:107.
6. Burkitts DP, Wright DH. Geographical Distribution of Burkitts Lymphoma. Edinburgh: E. & S. Livingstone, 1970;7:210.
7. Barber HRK, Graber E. Gynaecological tumours in childhood and adolescence. Obstet Gynaecol Survey 1973;28:357–81.
8. Nelson GA, Dockerty MD, Pratt JH, Remine WH. Malignant lymphoma involving the ovaries. Am J Obstet Gynaecol 1958;76:861–71.
9. Armon PJ. Burkitts lymphoma of the ovary in association with pregnancy. Two case reports. Br J Obstet Gynaecol 1976;83:169–72.
10. Williams AO. Tumours of childhood in Ibadan, Nig Cancer 1975;36:370–8.
11. McMahon B. Epidemiologic aspects of acute leukaemia and Burkitts tumour. Cancer 1968; 21:558–61.
12. Egwuatu VE, Ejeckam GC, Okaro JM. Burkitts Lymphoma of the vulva. Case report. Br J Obstet Gynaecol 1980;27:827–30.
13. Carbone PP, Berard CW, Bennett JM, Ziegler JL, Cohen MH, Gerber P. Burkitts Tumour. Ann Int Med 1969;70:817–32.
14. Burchenal JH. Long term survivors in acute leukaemia and Burkitts Tumour. Cancer 1968; 21:595–9.
15. Serov SF, Scully RE, Sobin IH. Histological Typing of Ovarian Tumours. Geneva: WHO 1973.
16. Woodruff JD, Noli Castello RD, Novak E. Lymphoma of the ovary. Am J Obstet Gynaecol 1973;85:912–18.
17. Zeigerman JH, Imbriglia J, Makler P, Smith JJ Ovarian lymphosarcoma, report of a case with review of literature. Am J Obstet Gynaecol 1956;72:1357–60.

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