Ovarian Teratomas Causing Gliomatosis Peritonei in Africans*

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SUMMARY

Two cases of gliomatosis peritonei in association with apparently benign ovarian teratomata are described. The patients were African children who are both alive and well, 4 years and 15 months respectively, following removal of the ovarian tumours.

It is suggested that cases of this type form a well defined, albeit rare, group with good prognosis and no attributable mortality. Accordingly, conservative management following removal of the primary tumour is advocated, especially with a view to the preservation of fertility.

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According to Fortt and Mathie,[1] only 13 cases of human gliomatosis peritonei associated with ovarian teratoma have been recorded since 1905: 4 other possible cases among the 11 listed by Proskauer,[2] are rejected by these authors because of uncertainty about the site of origin, outcome, or predominance of neural tissue in the peritoneal deposits. They also exclude the case of Luse and Vietti.[3]

This is an acount of 2 cases of this condition affecting African children. It has not previously been reported in Africans, although the second case of Berger and Pochaczevsky[4] was a Negro girl.

CASE REPORTS

Case 1

An 11-year-old African girl was admitted to Harari Hospital, Salisbury, Rhodesia, on 14 December 1966, having been referred from a district hospital.

She complained of a week-old swelling of the lower abdomen, mainly on the right side, accompanied by pain. She had also suffered from watery diarrhoea during the same period.

On inspection, the lower abdomen was distended (Fig. 1), and a large, firm, mobile, coarsely nodular mass could be felt in the right side of the abdomen extending from near the rib margin down into the right iliac fossa and across the midline in the suprapubic region into the left iliac fossa. The tumour was freely mobile laterally, but less so in the vertical direction. Tenderness was present. A fluid thrill was demonstrable.

*Date received: 14 June 1971.

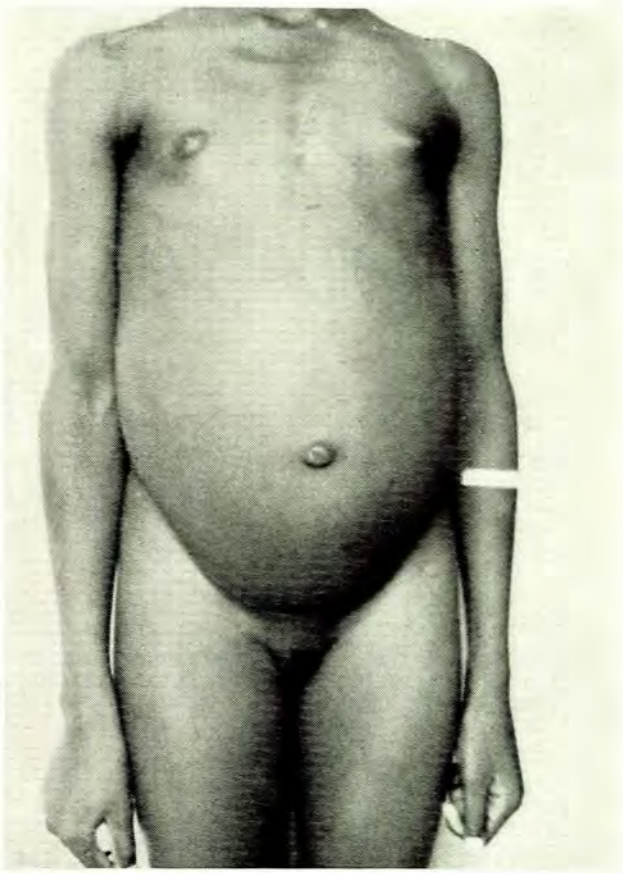

Fig. 1. Case 1. Bulging abdomen at time of admission.

Radiography of the abdomen showed rather diffuse calcification (Fig. 2). (Incidentally, radiography also showed fusion of the acetabulum and of the femoral head with the neck—a state appropriate to an age of 18 - 20 years.)

Blood urea was 33 mg/100 ml and intravenous pyelography did not show any abnormality. Haemoglobin was 10·2 g/100 ml and the faecal occult blood test was feebly positive. Hookworm ova were found in the stool. ESR was 50 mm in 1 hour (Westergren). No other abnormalities were found.

A pre-operative diagnosis of ovarian tumour was made and laparotomy and removal of the tumour were performed on 30 December 1966. Some ascitic fluid was noted at operation but the most striking additional

feature was the presence of numerous pale nodules, resembling miliary tubercles, studding the peritoneal surface, including the omentum (Fig. 3). The primary tumour arose from the right ovary, the other adnexae appearing normal.

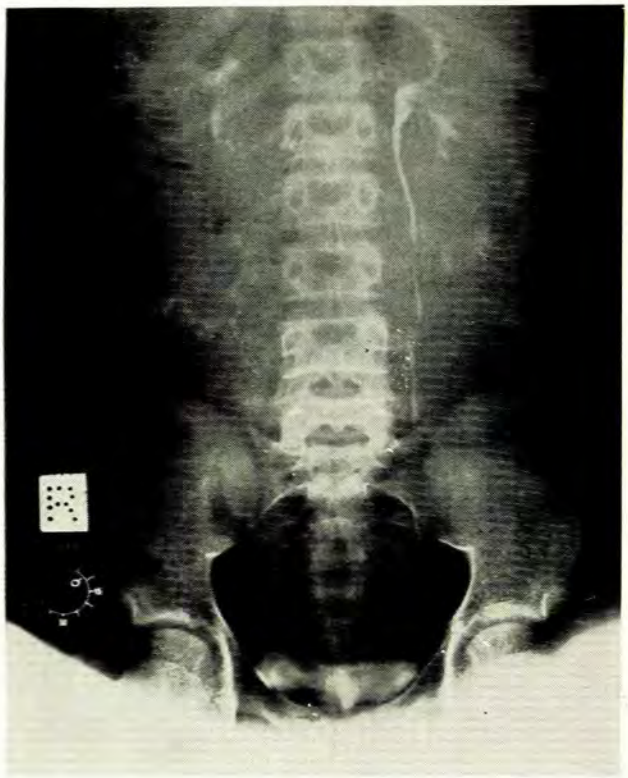

Fig. 2. Case 1. Intravenous pyelogram showing calcification in abdomen.

Fig. 3. Case 1. The tumour at operation. Note small nodules studding omentum to left of bowel loops.

The patient was initially treated with antituberculous chemotherapy which was discontinued following histological examination of the omentum. She made an uneventful recovery and was discharged from hospital on 14 January 1967.

Since discharge, this girl has been seen yearly, the last occasion being on 20 March 1971. At no time has any clinical abnormality been detected in her abdomen. Her menarche occurred in September 1969, and her menstrual cycle is normal and regular—4/28 days. She is now 16 years old but, as yet, has no marriage plans.

Case 2

A 6-year-old African girl was admitted to Nyadiri Mission Hospital, Rhodesia, on 5 October 1969, with a complaint of abdominal distension and shortness of breath for 1 year.

Her weight was 17·5 kg and the abdomen was tensely distended with ascitic fluid. Paracentesis abdominis yielded 3 litres of fluid and allowed palpation of a large left-sided abdominal mass. Her weight fell to 14·5 kg after paracentesis. Haemoglobin was 11·5 g/100 ml.

Laparotomy was performed on 3 November 1969. Little ascitic fluid was found. A tumour arose from the right ovarian pedicle. This was clamped and ligated, catching the top of the uterus. It was presumed that the fallopian tubes were also caught during this procedure. The tumour was removed. It was described as being 30 cm in diameter, with various cystic and solid areas which included skin, hair, bone and cartilage. Small nodules were noted studding the peritoneum and a sample of affected omentum was also taken.

Postoperative recovery was uneventful and the child was discharged from hospital on 18 November 1969, at which time she weighed 11·8 kg.

This girl was last seen on 13 February 1971. Apart from the paramedian scar extending from pubic crest to costal margin, and a small umbilical hernia, her abdomen was clinically normal and she was a healthy, happy child.

PATHOLOGY

The ovarian tumour from case 1 measured 28 × 20 × 10 cm, and weighed 3 200 g. On section, it was a partly cystic mass of irregular texture with very soft, pale tissue predominating. Bone and skin, bearing black hairs, were noted, together with mucinous accumulations.

On microscopy, the ovarian tumour showed a mixture of neural tissue, skin with appendages, smooth muscle, fat, bone and columnar epithelium, both mucous and ciliated. Ependymal structures and choroid plexus were identified in the neural portions but the bulk of this component had the appearance of glial tissue (Figs. 4 and 5).

The omental tissue showed the presence of numerous well-circumscribed nodules composed of fairly widely separated spindle cells with moderately fibrillar appearance and resembling the glial tissue of the ovarian tumour in

places (Figs. 6 and 7). In one respect, however, these nodules differed from the main tumour, in that sparse collagen fibres were present.

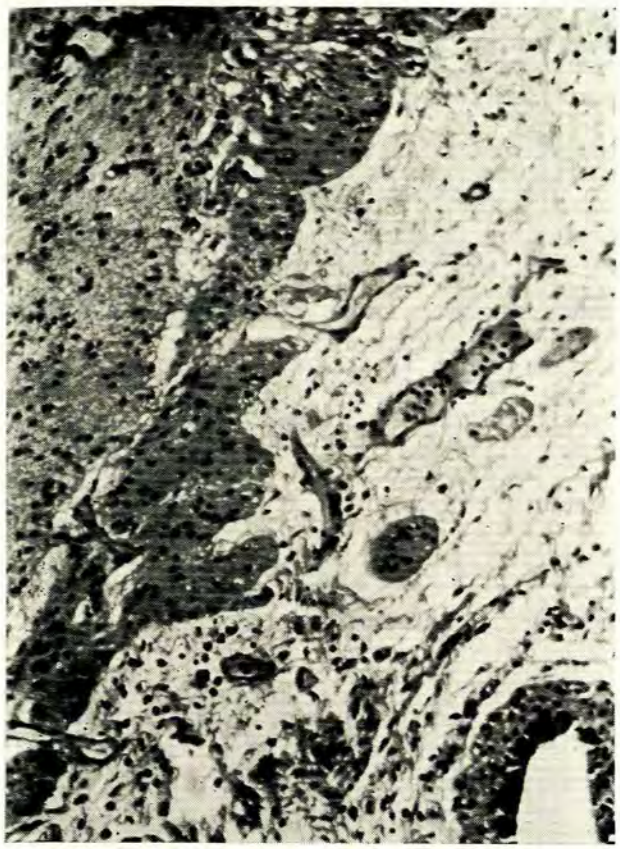

Fig. 4. Case 1. Section of primary tumour showing glial tissue and part of ependyma-lined cavity (H. and E. × 150).

Material from case 2 consisted only of samples of the ovarian tumour and a bit of omentum.

Microscopically, the presence of the tissues already noted were confirmed, with the additional demonstration of bronchial epithelium, muscle and collections of mildly whorled spindle cells. The omental deposits were similar to those seen in case 1, with the difference that collagen fibres were much more striking (Fig. 8).

DISCUSSION

Ovarian teratomas are not uncommon in Africans. In a 10-year survey of the histological material examined at Harari Hospital, these tumours were found to account for 21% of all ovarian tumours.

Willis[5] has categorized the malignant or metastatic propensities of teratomas into 3 types:

(i) those in which the whole tumour is malignant,

(ii) those in which only one component becomes malignant, and

(iii) those in which peritoneal dissemination of relatively benign tissue occurs.

These 2 cases belong in the last category, which is the rarest form of spread. So benign was the appearance of the peritoneal deposits that it seems improbable that they could be the result of truly malignant dissemination. Helmke[6] interpreted this condition as being one of benign tissue transplantation and Proskauer[2] referred to implantation metastases. Noguchi and Lonser[7] postulated that spillage from an earlier biopsy was responsible for seeding of the tumour in their case and suggested that the peritoneum has a special affinity for glial cells. Fortt and Mathie[1] suggested that the prognosis in this condition is fairly good when only mature tissues appear in the primary tumour and in the peritoneal deposits.

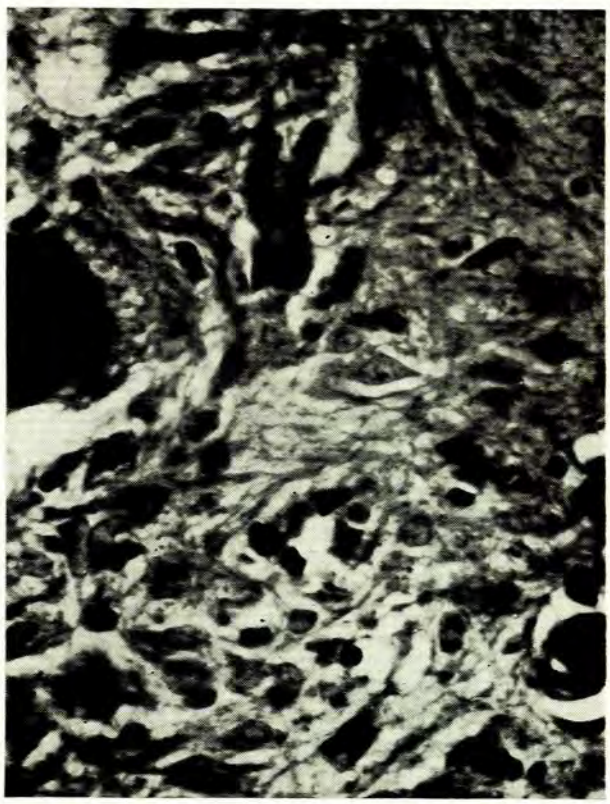

Fig. 5. Case 1. Glial tissue in primary tumour (Heidenhain × 750).

In this connection, it is possible to reassess the lists of reported cases. The fatal cases listed by Proskauer,[2] but excluded by Fortt and Mathie,[1] all deviated from the pathological picture of gliomatosis peritonei by showing such features as 'embryonal structure', 'gland-like proliferations' or 'resemblance to round-cell sarcoma or glioblastoma'. Among the cases tabled by Fortt and Mathie,[1] those of Schairer[8] and Benirschke *et al.*[9] were both rapidly fatal and did not show purely glial peritoneal deposits. Neuhäuser's[10] case was not fatal within 8 months

but was described as resembling round-cell sarcoma and should, therefore, be omitted. Those cases in which the peritoneal deposits consisted only of well-differentiated glial tissue number a mere dozen (Table I).

The most significant feature of this table is that it shows no resultant deaths. It is evident that the prognosis is, in fact, very good. The most notable survival is that of the patient of Thurlbeck and Scully,[11] who has survived at least 26 years since the diagnosis was made. Other long survivals include the case of Ackerman[12] and the first case in this account.

There arises the question of the fate of the peritoneal nodules in these cases. In the two examples here presented, the presence of collagen and the general appearance of the deposits were considered very reminiscent of neurofibromatosis. Such a resemblance could be a result of Schwann cells fulfilling the role of facultative fibroblasts.[13] Schwann cells are closely similar to neuroglia and also arise, normally, from the neural crest. It can, therefore, be postulated that the peritoneal deposits may behave much like neurofibromata and carry no greater risk of malignant degeneration. The alternative possibility of peritoneal fibromatosis coincidental with ovarian teratoma appears unlikely.

TABLE I. REPORTED CASES OF GLIOMATOSIS PERITONEI WITH OVARIAN TERATOMAS

Case report	Patient's age	Outcome
Büttner (1909)[14]	20 years	Alive and well 3 years later
Nordmark (1932)[15]	11 years	Alive and well 18 months later
Helmke (1938)[6]	16 years	Alive and well 1 year later
Proskauer (1946)[2]	22 years	Alive and well 15 months later
Thurlbeck and Scully (1960)[11]	18 years	Alive and well 26 years later
Noguchi and Lonser (1961)[7]	22 months	Alive and well 2 years later
Kourie and Roujeau (1966)[16]	9 years	Alive and well 1 year later
Ackerman (1968)[12]	Child	Alive and well 5 years later
Fortt and Mathie (1969)[1]	18 years	Alive and well 18 months later
Berger and Pochaczevsky (1969)[4]	12 years	Alive and well 1 year later
Present case 1.	11 years	Alive and well over 4 years later
Present case 2.	6 years	Alive and well 15 months later

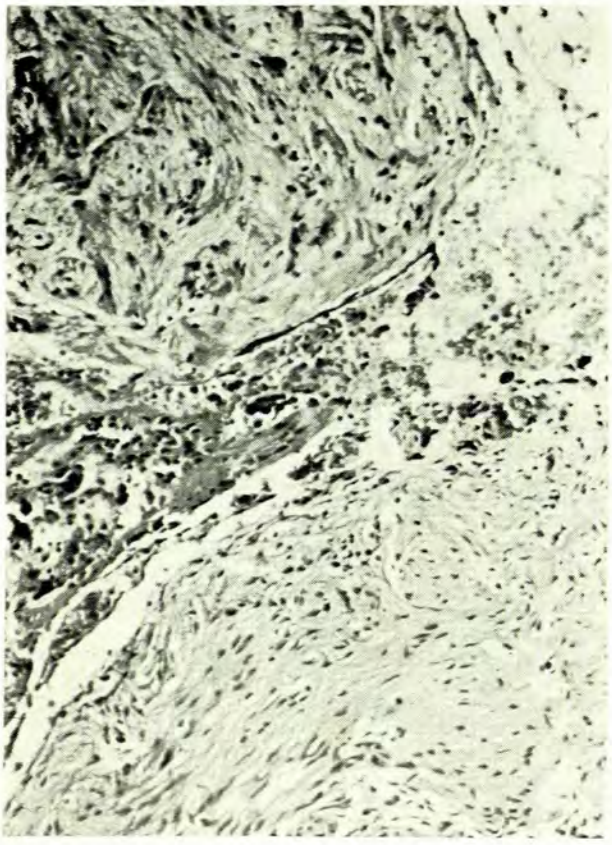

Fig. 6. Case 1. Section of peritoneal deposit (H. and E. × 150).

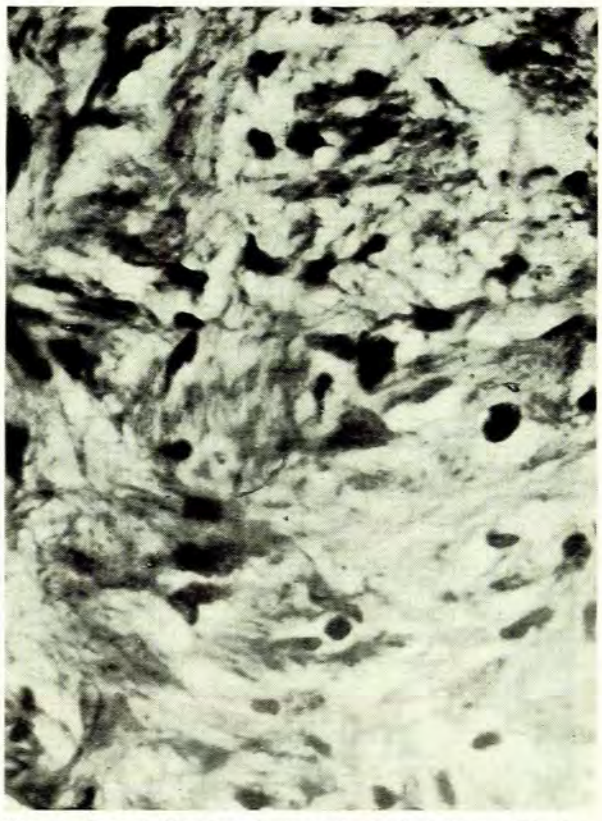

Fig. 7. Case 1. Peritoneal deposit of glial tissue (Heidenhain × 750).

Proskauer[2] has suggested that these metastatic nodules may undergo regressive changes after removal of the primary tumour. Having regard to the structure of these nodules, this seems unlikely. The second case of Berger and Pochaczevsky[4] was subjected to a 'second look' operation a year after removal of the teratoma when the peritoneal deposits were found to be apparently unchanged.

Fig. 8. Case 2. Peritoneal deposit showing collagen fibres (Van Gieson × 190).

In view of these aspects, it is doubtful whether radiotherapy can make a useful contribution to the management of this condition. Although they employed it, Thurlbeck and Scully[11] considered that evidence of benefit was very weak. As the victims of this condition are all young (ages ranged from 22 months to 22 years), irradiation must be deemed undesirable as well as redundant.

In so far as both the cases here presented were spared from exposure to irradiation, fertility may be preserved. The first patient should soon be marrying and, as contact has been maintained, there is hope that she may become the first recorded victim of this condition to achieve subsequent motherhood. The prospects of the second girl are more dubious but it may be that the unaffected fallopian tube and ovary were spared at operation.

We should like to thank Professor J. H. Mynors and Dr J. Buterbaugh, for their assistance and for allowing us to report these cases. We also wish to thank Drs R. Johnson and E. Mackintosh, Professor R. H. Philpott and Mr G. Davies for their help; and Dr E. Burnett-Smith, Deputy Secretary for Health, for permission to publish.

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