Case Report

ARRHENOBLASTOMA

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The case of a patient with an arrhenoblastoma who presented as an acute abdominal emergency, is reported here.

Arrhenoblastoma is a rare, predominantly unilateral, ovarian tumour of mesenchymal origin that causes defeminizing and striking masculinizing effects in the female. It usually occurs in the child-bearing age group and its malignant potential is estimated at approximately 25%. This case is presented because of the rarity of this fascinating tumour and especially because it constituted an acute abdominal emergency.

BRIEF REVIEW OF THE LITERATURE

Pick is given credit for describing the first case in 1905. Javert and Finn, however, cite the case of Davis in 1900, of clinical virilization accompanying an ovarian tumour, as one of an arrhenoblastoma. In 1931 Meyer suggested the name 'arrhenoblastoma' when reporting his experience with 26 cases and also described the three main histological types: (1) The well-differentiated tubular adenoma, (2) the intermediate type, and (3) the undifferentiated sarcomatoid variety. He further proposed an origin from male-directed cells situated in the region of the hilum. In 1940 Kanter and Klawans reported the largest arrhenoblastoma: it measured 28 cm. in diameter and weighed 4,640 G. During the past 2 decades several reports, pertaining to incidence, histogenetic theories, variation of gross and microscopic appearances and malignant potential, have appeared. Following Iverson's review in 1947, Javert and Finn described a 25% malignant potential in their review. After an all-inclusive discussion Hughesdon and Frasier suggested an origin from ovarian stroma directly, thus constituting 'a mixed mesodermal tumour with coelomic, testicular and Müllerian derivatives'. More recent reviews have been reported by Pedowitz and O'Brian, Hertig and Gore, and Fontana and Simpson. From all these reports it is clear that the tumour varies greatly in its size, shape and gross appearances, and about 5% are bilateral. Most authors recommended radical surgical treatment. Javert and Finn suggest conservative therapy, such as unilateral oophorectomy—only in the young female who desires more pregnancies and is stable in weight.

CASE REPORT

The patient was a 36-year-old Negro, gravida 5, para 5, and she was divorced. Her youngest child was 9 years old. She was admitted during the night of 22 March 1963 with a history of sudden onset of acute abdominal pain in the lower abdomen

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which radiated to the tip of her right shoulder. The pain was sharp, severe and persistent and was accompanied by dizziness and nausea. There was no vaginal bleeding or history of trauma. Her last menstrual period was at the beginning of January 1963 and lasted 4 days. Physical examination revealed a female in severe distress. She was of normal stature and body contour. Pulse rate 104/min., blood pressure 90/70 mm.Hg, temperature 98·6°F, respiration 30/min. Her skin was warm but moist. Abdominal examination revealed slight distension of the lower abdomen with marked guarding, rigidity and bilateral rebound tenderness. On pelvic examination the external genitalia appeared normal. She had a marital introitus and a normal vagina. The cervix was firm, pink, very tender on motion, and the os was closed. There was a slight fullness in the *cul de sac* with a vague, tender mass in the region of the right adnexa. The left adnexa and uterus could not be well delineated because of the extreme tenderness.

The haematocrit was 32% and the urine was negative. The pre-operative diagnosis of a ruptured tubal gestation was made, and after blood transfusions had been commenced an immediate laparotomy was performed. A large amount of fresh blood and clots (estimated at 1,500 ml.) was encountered in the peritoneal cavity. The left adnexa and uterus were normal. The right fallopian tube was normal, but the right ovary was enlarged (15 × 5 cm.) and was firm, pinkish-yellow in its medial half, while the lateral half was reddish, rubbery, and had ruptured on the posterior aspect where it was bleeding actively. A right salpingo-oophorectomy was performed. Reddish, rubbery, friable tissue, not unlike placental tissue, could be scooped out of the lateral half of the ovarian mass, and a presumptive postoperative diagnosis of a ruptured ovarian pregnancy was made. The patient's postoperative course was completely uneventful.

Pathological Findings

The gross specimen measured 15 cm. in length and was 5 cm. in width at its widest part. It was rubbery in consistency and slightly lobulated. The outer surface was reddish in colour and at the point of perforation dark red. The cut surface had a homogeneous yellow appearance except for the dark red portion where haemorrhage had occurred. There were no cysts present. Microscopic examination showed hyperchromatic epithelial cells arranged in acini and tortuous cord formation interspersed with occasional pale staining cells. There was no actual tubular formation but merely a suggestion of imperfectly formed glandular tissue. A high-power view showed that the hyperchromatic nuclei varied greatly in size and shape, from small round to large spindle-shaped nuclei, but nevertheless arranged in cords and acini formation. There is just a suggestion of luteinization of the mesenchymal stroma (Fig. 1).

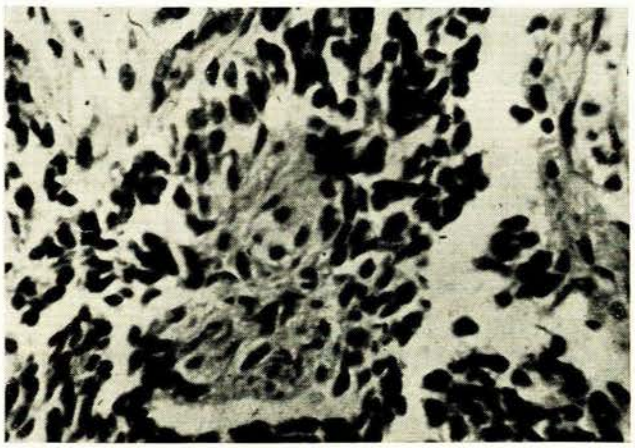

Fig. 1. See text.

This microscopic pattern was repeated in all sections taken from the tumour, even those taken from the site where perforation had occurred and from the tissue that was scooped out at the time of laparotomy. The fallopian tube was completely normal. The pathological diagnosis was arrhenoblastoma of the intermediate variety. Subsequent history revealed the following salient points:

1. Although she bled for 4 days in January 1963, she had been amenorrhoeic since 1958 and occasionally skipped periods since 1955.

2. Hirsutism had been present for 7 years, and she had shaved a few hours before the sudden onset of pain. Besides shaving regularly she had tried electrolysis therapy to no avail.

3. Acne on the face and trunk had been present for 3 years.

4. Up until 1955 she could sing very well. From 1955 to 1958 she noticed that she no longer could sing as before. In 1958 hoarseness of the voice began and then persisted.

5. She had been told of a 'fibroid tumour' on routine examination in 1955 for which she received injections to 'shrink the tumour'. She gave up the injections after 3 years owing to lack of funds.

COMMENT

It is most unfortunate that this patient did not seek proper medical advice for her symptoms in 1958 or soon thereafter. One can merely speculate what might have occurred had the tumour not ruptured. In fact, the perforation may well turn out to be a blessing in disguise. It is interesting that despite this patient's masculinizing features her clitoris and her breasts remained normal in size. Furthermore, it is no less interesting that she should have had 4 days of vaginal bleeding after 5 years of amenorrhoea. Fluctuating circulating oestrogen levels could possibly account for this. Her vaginal smear showed moderate oestrogen effect. Although the history was of 8 years' duration and the tumour was macroscopically unilateral and of the intermediate type, we felt that because it had ruptured, this was an indication of fairly sudden growth and that it therefore warranted more radical surgery. In addition we recommended that, if an unsuspected solid ovarian tumour is encountered at the time of laparotomy, a frozen section should be requested in order that the correct surgical treatment could be instituted forthwith, provided the patient's general condition was satisfactory. The problem was discussed and explained to the patient in detail, but she refused further surgical treatment, expressing the hope of a second marriage and possibly more children.

Follow up. The patient began menstruating regularly 2 months after surgery. The hoarseness, acne and hirsutism, although less severe, have persisted. To date there has been no clinical or radiological evidence of recurrence or metastasis.

SUMMARY

1. A brief review of the literature describing arrhenoblastoma is presented.

2. An additional case is reported, the presenting symptom of the patient being an acute abdominal emergency.

3. The interesting facets in this case are commented upon.

4. Choice of treatment is discussed and conservative therapy is only recommended in the young patient who desires further children and in whom the tumour is benign and unilateral.

5. It is suggested that a frozen section should always be requested when an unsuspected solid ovarian tumour is encountered at laparotomy.

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