Granulosa Cell Tumour: A Recurrence 40 Years After Initial Diagnosis

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Abstract

Background: Granulosa cell tumours are rare ovarian malignancies accounting for 2% to 5% of ovarian tumours. They are characterized by late recurrences, with the latest recurrence reported 37 years after first diagnosis.

Case: A 73-year-old woman who had surgery for a granulosa cell tumour in her early 30s and who presented with a recurrence 40 years later.

Conclusion: Patients and their physicians should be made aware that there is a potential for a granulosa cell tumour to recur many years in the future.

Résumé

Contexte : Les tumeurs de la granulosa sont des tumeurs malignes ovariennes rares qui représentent de 2 % à 5 % de l'ensemble des tumeurs ovariennes. Elles sont caractérisées par des rechutes tardives, la rechute la plus tardive ayant été signalée 37 ans après le diagnostic d'origine.

Cas : Une femme de 73 ans qui avait subi, au début de la trentaine, une chirurgie en raison d'une tumeur de la granulosa et qui a présenté une rechute 40 ans plus tard.

Conclusion : Les patientes et leurs médecins devraient être mis au courant du potentiel de rechute, de nombreuses années plus tard, des tumeurs de la granulosa.

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INTRODUCTION

Granulosa cell tumours are rare ovarian malignancies accounting for 2% to 5% of ovarian tumours.[1,2] Most granulosa cell tumours present at Stage I. Although some may behave aggressively, most have a low malignant potential and evolve slowly, with the natural course of the disease taking many years.[1] Patients usually have a good prognosis, with a 5-year survival of up to 90%.[1] Recurrences usually are identified at a mean interval of 6 years after initial presentation.[1] Here we report a case of disease recurrence 40 years after the initial presentation, which, to our knowledge, is the longest interval from initial presentation to recurrence yet reported.

Key Words: Granulosa cell tumour, recurrence

Competing interests: None declared.

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CASE

Christine (pseudonym), a 73-year-old nulliparous woman, presented with abdominal discomfort of 1 year's duration. During this time, she had weight loss of approximately 9 kg. Initial investigations included a CT scan, which demonstrated a mixed-density mass, 3.8 × 1.5 cm in diameter, close to the anterior aspect of the bladder. Since the nature of the mass was unknown, Christine underwent a CT-guided biopsy (Figure 1).

The pathology report indicated that the tissue was consistent with a recurrent granulosa cell tumour (Figure 2). The strong inhibin marking on immunohistochemistry was consistent with the diagnosis.

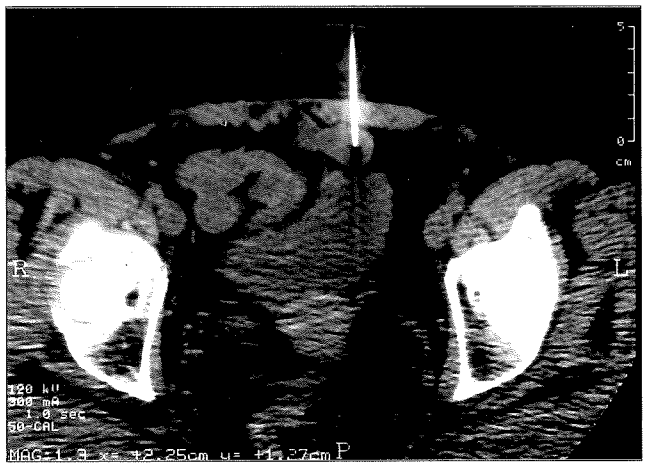
Christine's medical history included undergoing a left salpingo-oophorectomy for a granulosa cell tumour at age 33 years. She did not receive any adjuvant therapy and was subsequently lost to follow-up.

Christine was referred to our centre in February 2002 after the result of the biopsy was reported. She had vaginal bleeding at the time of referral. An endometrial biopsy was attempted but could not be completed because of cervical stenosis. Pelvic examination revealed a small, retroflexed uterus with no palpable adnexal masses. The serum level of CA-125 was normal (16 KU/L), and serum inhibin B was mildly elevated (81 ng/L).

Christine underwent an exploratory laparotomy in March 2002. During surgery, the pelvis, right ovary, and uterus were normal, as was the exploration of the upper abdomen. No suspicious lymph nodes were identified. A 3-cm diameter mass, located in the space of Retzius, was excised, and a total abdominal hysterectomy and right salpingo-oophorectomy was performed.

The pathology report indicated that the pelvic mass was consistent with recurrent granulosa cell tumour. The tumour cells were positive for vimentine, alpha-inhibin, and progesterone receptors. The right ovary and tube were

Figure 1. CT-scan guided biopsy of the pelvic mass



normal. The endometrium showed atypical hyperplasia and a polyp with cellular atypia.

Christine received no adjuvant treatment, and her postoperative recovery was uneventful. At her most recent follow-up visit, 18 months after surgery, Christine continued to do well, with no evidence of recurrence. Her serum inhibin level was 59 ng/L and serum CA-125 was 11 KU/L (both normal values).

Christine died 30 months after her surgery from a myocardial infarction.

DISCUSSION

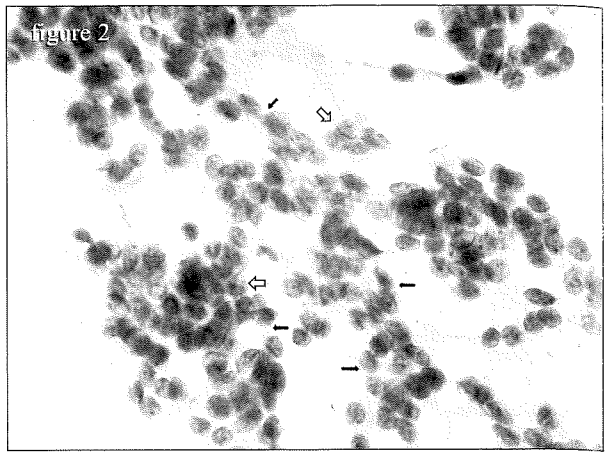
The 2 different types of granulosa cell tumour are the juvenile and the adult type. The adult type is more common, and the mean age of presentation is 50 to 54 years. These tumours may be hormonally active, producing estrogens and, rarely, androgens.[1]

As most series are small, there is no definitive consensus on ideal management. Surgical removal of the tumour is the mainstay of management.[1–3] Conservative surgery has been advocated for the group of women who may wish to preserve their fertility potential. Granulosa cell tumours are rarely bilateral, and they are usually confined to the ovary.[1]

In women who do not require preservation of fertility and in those with more advanced disease, total abdominal hysterectomy and bilateral salpingo-oophorectomy is the treatment of choice.[3] Because these tumours have a slow growth pattern, approximately 90% of women present with Stage I disease at diagnosis.[3] Recurrences have been documented in 9% of cases and are usually local. The tumours may recur late; the mean time between diagnosis and recurrence is 4 to 6 years,[1,3] but recurrences at between 13 and 36 years have been described.[4,5] Hines reported a recurrence 37 years after the first episode.[2]

Treatment for patients with a late recurrence of the tumour is an attempt at surgical excision. Postoperative adjuvant therapy may be considered.[1–3]

Figure 2. Membrane irregularities or grooves (white arrows) and Call-Exner bodies (black arrows) on cytology. HPS staining. Magnification: X20.



The woman whose case is described here had a granulosa cell tumour of the left ovary, removed 40 years prior to her presentation. Since the right ovary was normal on pathological review, we believe that the resected mass represented a pelvic recurrence of her primary tumour, the most common site of recurrence.[3]

To our knowledge, this case represents the longest interval from initial presentation of granulosa cell tumour to recurrence yet reported.

CONCLUSION

Women treated for granulosa cell tumour may have late recurrences despite having a prolonged disease-free interval. Patients and their physicians should be made aware of the potential of these tumours to recur very late.

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The next-of-kin of the woman whose story is told in this case report has provided signed permission for its publication.

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