CASE REPORT

Combined Ganglioneuroma and Endometriosis of the Vaginal Fornix.

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Ganglioneuromas are relatively rare neoplasms occurring in the mediastinum, retroperitoneum and adrenal glands and occasionally in the cervix and fallopian tube. Conversely, endometriosis is a relatively common condition and can occur in multiple sites. However, endometriosis in association with ganglioneuroma has not been documented. Here, we describe a case of ganglioneuroma and endometriosis in the vaginal fornix.

INTRODUCTION

Ganglioneuromas are relatively rare neoplasms occurring in decreasing order of frequency in the mediastinum, retroperitoneum and adrenal glands (1). Occasionally, ganglioneuromas have been reported in the cervix (2) and in the fallopian tube (3). Ganglioneuromas have not been reported in the vagina. Endometriosis, on the other hand, is a relatively common condition and can occur in multiple sites, including the vagina (1), but its association with neural tumors has not been documented.

This report pertains to a case of ganglioneuroma and endometriosis in the vaginal fornix.

CASE HISTORY

A 32 year old white woman was admitted to the Victoria General Hospital with complaints of menorrhagia and dyspareunia. Physical examination showed a multiparous cervix with no obvious lesions and no adnexal masses. A 3.0 centimetre tender nodule was found in the right lateral vaginal fornix. The remainder of the physical exam was unremarkable.

Relevant past history included three pregnancies with uncomplicated spontaneous vaginal deliveries and the use of an intrauterine contraceptive device six years prior to admission. A fine needle aspirate was diagnosed as "likely" endometrioma. Family and social history were noncontributory. All the laboratory tests were found to be within normal range. An abdominal hysterectomy with bilateral salpingo-oophorectomy and excision of a nodule on the right vaginal fornix were performed. The post-operative course was unremarkable and the patient was discharged seven days post-surgery.

Samples from the nodule on the right vaginal fornix were studied by light microscopy after formalin fixation. Sections were stained by hematoxylin and eosin, Masson's trichrome and Bielschowsky's silver impregnation. Immunoperoxidase for protein S-100 was also performed on formalin fixed tissue. Samples from the hysterectomy specimen were studied with hematoxylin and eosin.

PATHOLOGICAL FINDINGS

The uterus was grossly unremarkable. The ovaries had small follicular cysts and corpora lutea. The vaginal fornix specimen consisted of a strip of tissue lin-

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ed by normally appearing vaginal mucosa. It measured 8 x 3 x 0.4 centimetres. On the deeper aspect, there was an ill-defined nodular structure measuring 3.5 x 3 x 3 centimetres. The cut surface revealed a variegated, poorly demarcated fibrous lesion.

Microscopic examination of the uterus revealed foci of superficial adenomyosis. The ovaries had bilateral follicular cysts and the fallopian tubes were unremarkable.

Sections from the vaginal fornix showed normal vaginal epithelium. Deep in the vaginal stroma, there was an ill-defined lesion consisting of bundles of nerve fibres and Schwann cells, as well as numerous ganglion cells (Figure 1). No satellite cells were identified and the ganglion cells lacked Nissl granules. A second component of this lesion consisted of islets of endometrial tissue containing both glands and stroma. Although there were islets consisting solely of endometrial elements, in most instances both endometrial and neural elements were intimately associated (Figure 2). Gan-

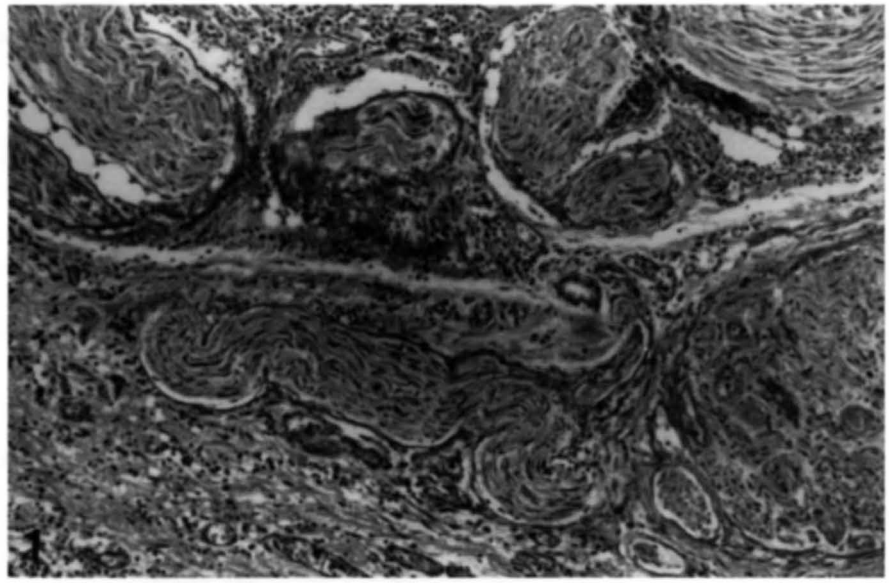

Figure 1: *Vaginal stromal lesion consisting of disarrayed bundles of nerve fibres, Schwann cells and ganglion cells. (Hematoxylin-eosin, X150)*

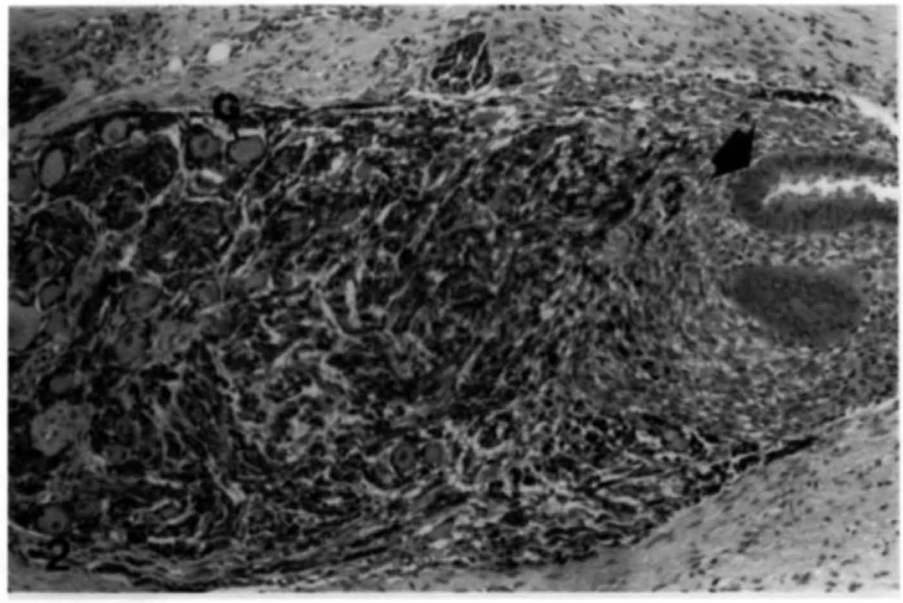

Figure 2: *Vaginal stroma showing endometrial tissue inclusions (arrow) admixed with neural elements including ganglion cells (G). (Hematoxylin-eosin, X150)*

glion cells were often surrounded by endometrial stromal elements (Figure 3).

DISCUSSION

Ganglioneuromas are neoplasms composed of nerve fibres, Schwann cells and ganglion cells at various stages of maturity. Ganglioneuromas can occur in many locations but are most commonly found in the sympathetic ganglia, especially the posterior mediastinum and retroperitoneum (2). Less common sites associated with smaller autonomic ganglia have also been reported (4). Ganglioneuromas predominate in females in a ratio of 3:2 over males (4). They are found most often before the age of 20 years, but have been described in elderly patients. The most common presentation results from its mass effect on adjacent structures resulting in pain or discomfort. However, a large number of these tumors are discovered as asymptomatic masses.

Ganglioneuroma associated with endometriosis has not been previously described. The occasional presence of neural tissue, specifically glial tissue, in the uterus (5-8) has been explained as resulting from implantation of fetal tissue following spontaneous abortion, miscarriage or curettage for therapeutic abortion (5,6,8). Neurofibromas in the female organs have been described in patients with neurofibromatosis (9). Endometriosis with perineural involvement has been described and felt to be due to expansion of the endometrial tissue into a tissue plane of least resistance, the perineural space (10). In a report of ganglioneuroma in the cervix, Fingerland *et al.* (2) mentioned the presence of endometrial tissue within the ganglioneuroma but this occurred in an endometrial polyp without endometriosis.

Treatment of ganglioneuromas is by complete surgical removal. This may be difficult or even impossible if the tumor is very large, ill-defined and/or adherent to adjacent structures. With complete removal, recurrence should not occur.

While endometriosis is relatively common, it is rarely associated with neoplasms. A rare case of a sarcoma associated with omental endometriosis was reported by Coutiforis *et al.* (11) and they elaborated on the possibility that adenocarcinoma or sarcoma may develop on a substrate of endometriosis.

The present case of a plexiform ganglioneuroma associated with endometriosis is likely a fortuitous, albeit intriguing, heterotopic occurrence. This patient had three normal vaginal deliveries and therefore no history suggestive of traumatic implantation of fetal tissue.

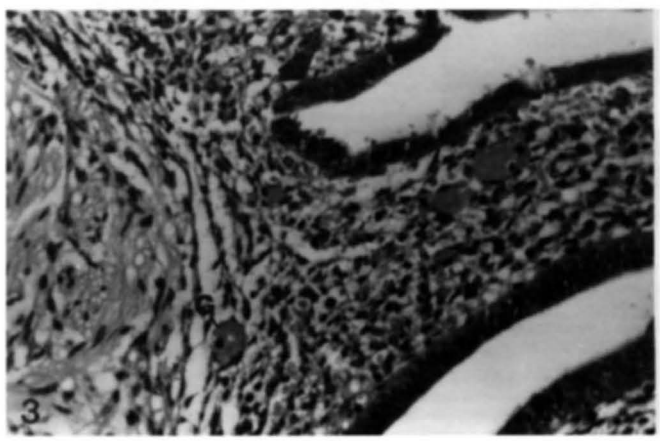

Figure 3: *Vaginal stroma (higher magnification) depicting the close relationship between endometrial (arrow) and neural tissue (G). (Hematoxylin-eosin, X450)*

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