Case title: The Case of Mrs. Virginia Jones*

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Case synopsis: A 50 year old woman is referred to her gynecologist for the

assessment & management of her chronic vulvar itching. She is lost to follow up and returns with a vulvar lesion.

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Case objectives: As part of an adult learning experience the learners should

identify what they need to know to solve this patient's case. The learning goal of this case is to facilitate the clinical

reasoning skills of the learners.

The authors used published objectives from the Association

of Professors of Obstetrics & Gynaecology of Canada (APOG), Association of Professors of Gynecology and Obstetrics (APGO) and Council of Residency Education of Obstetrics & Gynecology (CREOG) as guidance for writing

these cases.

This case was designed for a small group interactive 2.5 hour seminar – the case may be revised as needed into 2

parts to fit a shorter time frame.

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You are a gynaecology resident on a 3 month winter "ambulatory community rotation" with Dr. Ski - a gynaecologist in Whistler, BC. The next patient is a Mrs. Virginia Smith - a 50 year old retired criminal lawyer.

She has been referred by her family physician. Mrs Jones complains of a long history (>10 years) of itching of the vulvar skin. She changed family doctors recently and her new family doctor completed a physical exam 1 month ago. The physician was concerned because she noticed the vulvar anatomy was "abnormal" and the skin was white and has referred her for further assessment.

Dr. Ski asks you

"What is your differential diagnosis for vulvar itch?" "What other information would you like from her history?"

1. Address Dr. Ski's questions.

Mrs. Jones states she has "always" had mild to moderate itching involving the skin of her vulva and around her anus. She has seen numerous physicians about the problem and was treated for years for "recurring yeast infections". Eventually was told she had "pruritis vulvae and haemorrhoidal dermatitis". She was given a hydrocortisone cream to use when it "flares up". The cream works but as soon as she stops using the cream the itch comes back. She has not used the cream for years – "I guess I have just gotten used to it".

She denies any other skin, hair and or nail problems. She does not use scented soaps, perfumes, or powders in that area. Through out her 20's she also had problems with recurring yeast infections which she treated with anti-yeast vaginal medications. She doesn't think that yeast is the problem now since none of the over the counter anti-yeast medications help her itching.

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She is unsure how long the whitening of the vulvar skin has been present because "I've never looked at that skin and no one has ever mentioned it before".

She is a Gravida 2 Parity 2 – pregnancies, labour and deliveries unremarkable. She had a cone biopsy done in her 30's for carcinoma in situ of the cervix. All her Pap smears since the cone biopsy have been normal. Her last Pap smear was a couple of years ago. She stopped menstruating when she was 50 and has not had any vaginal bleeding since then. She is not interested in taking any hormone replacement therapy because of all of the "controversy". Her family doctor wanted to start her on topical estrogen therapy for her vulvar skin but she refused. She is an otherwise healthy 60 year old woman without any medical problems. She is currently not on any medications and denies any allergies.

- 1. Given this information, what is your differential?
- 2. What is the most likely diagnosis at this point?
- 3. Describe how you would perform the physical examination?

A general survey of the skin of the rest of the body does not reveal any abnormalities. On pelvic examination, inspection of the genitalia reveals abnormal vulvar anatomy. In particular, the top of the glans of the clitoris is just visible and the hood of the clitoris can not be retracted, there is shrinkage of the labia minora, and narrowing of the introitus. There is mild perianal inflammation. All of the skin of the vulva appears thin and atrophic. There are superficial excoriations noted on the medial aspects of the labia majora. (Figure 1). A speculum exam reveals a red flattened vagina. Inspection of the cervix reveals an unsatisfactory but negative colposcopic examination. Minimal white vaginal discharge is noted. The pH of the discharge is 5.0. A bimanual reveals a small mobile anterverted anteflexed uterus with no adnexal masses palpable.

- 1. Given this information from her physical examination, what is the most likely diagnosis now?
- 2. What investigations, if any, would you do at this point?
- 3. What is the next step in the management of this patient?

The patient refuses to have a vulvar skin biopsy done. Given the most likely clinical diagnosis, Dr. Ski agrees to treat the patient for 4 weeks and then reassess her condition. She understands that if there is no improvement over the interval of time she will need to have a skin biopsy at that point.

Dr. Ski leaves you to explain what the diagnosis is to the patient and answer any of her questions and write out a prescription for her medication.

- 1. Explain the diagnosis to the patient
- 2. What are your recommendations regarding treatment? (Be specific regarding dose, frequency and duration of therapy and follow-up).
- 3. Will topical therapy affect the results of a subsequent skin biopsy? If so, how?

Nine months later you have graduated and are doing a locum for Dr. Ski.

Mrs. Jones returns to the clinic for her "follow-up" visit. She tells you she suffers from seasonal affective disorder and following her last appointment with you "slipped into a bad depression". She did not use any of the medication you had prescribed.

Over the last 6 weeks she has noticed a painful lesion on the inside of her left labia majus that "just will not heal". It hurts when she urinates. On inspection the distortion to the vulvar anatomy is more pronounced the labia minora are completely gone. The glans of the clitoris is completely buried. There is marked inflammation and atrophy of the entire vulva and peri-anal skin area. There is a 1 x 1 cm lesion on the inside of the left labia majus. Her groin nodes are not palpable. (Figure 2)

She asks you "Is it the same thing going on?"

You recommend a skin biopsy, you mention there are a number of things that you can do to make it less painful, and this time she agrees.

- 1. What is the differential diagnosis?
- 2. Describe how you would perform a skin biopsy

You schedule a follow-up appointment for Mrs. Jones in 1 week. The biopsy report returns with the following diagnosis; "changes compatible with a high grade squamous intra-epithelial lesion in a background of lichen sclerosus". Refer to Figure 3

The patient asks you

"What does the biopsy mean? Do I have cancer?"

"How did this happen, I thought I just had a skin condition?"

"Will it go away if I use the steroid cream?"

You recommend a wide local excision for definitive diagnosis.

1. Address the patient's questions

Two weeks later you perform a partial vulvectomy. The pathology report is received a week later and reports "A well differentiated non keratinizing squamous cell carcinoma measuring 1 cm in greatest diameter and maximum 1 mm stromal invastion is noted in the center of the specimen. The surgical margins are uninvolved with the distance of the invasive carcinoma from the closest margin 1 cm. There is no obvious lymphvascular involvement. Extensive lichen sclerosus in noted in the remainder of the specimen." Now you have the pleasure of having George, a keen 3rd year obstetrics/gynaecology resident, working with you on a 3 month elective and asking them questions. You ask the resident ...

"How does vulvar cancer spread?"

"How is vulvar cancer staged?

"What is this patient's risk of having nodal involvement?"

"Does she need any further investigations and or treatment?

"What is her prognosis?"

1. What responses are you looking for?