

Granular Cell Tumor of the Vulva

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Granular cell tumors are rare soft-tissue neoplasms of neural origin. They usually occur in the head and neck region; however, up to 15% occur in the vulvar area.¹ The tumors are usually benign; however, malignant tumors have

been reported. Although uncommon, granular cell tumors should be included in the differential diagnosis of nodular vulvar lesions.

CASE REPORT

A 22-year-old white woman gravida zero presented to the office with a 4-year history of a “bump” on her vulva (Figure 1). The area was not painful, but she complained that her underwear would stick to the area. She had a history of chlamydia infection but was otherwise healthy. Testing was negative for gonorrhea, rapid plasma reagin, and HIV, and positive for chlamydia, for which she was treated with azithromycin. Punch biopsy of the area revealed a granular cell tumor.

The patient was then scheduled for surgery, and a wide local excision of the tumor was performed (Figure 2). The area healed, and she is doing well postoperatively. The pathology report confirmed the granular cell tumor.

DISCUSSION

Granular cell tumors are soft-tissue neoplasms of nerve sheath origin. The tumors may occur in children or adults but are most common in the third and fourth decade of life.¹ They are also seen more often in females than males and more frequently in African Americans than Caucasians.² The female-to-male ratio is approximately 3 to 2.³ Although they can occur anywhere on the body, more than half are found in the head and neck area, especially the tongue. The tongue is involved in 25% of cases.³

Granular cell tumors have been reported in the breast, gastrointestinal tract, cervix,



FIGURE 1. Granular cell tumor of the vulva.
Photos courtesy of Barbara Melican Gleason, DO.



FIGURE 2. Wide local excision of tumor.

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lungs, parotid, penis, scrotum, peripheral nerves, skin, and vulva.⁴ They have also been reported in the eyelid and appendix.⁵ It is estimated that between 5% and 16% occur in the vulva.⁶ Five percent of granular cell tumors are found in the gastrointestinal tract; if they are large enough there, they may cause symptoms of pressure or obstruction.³

Granular cell tumors usually present as nontender nodules and are typically smaller than 3 cm. A nodular mass may be seen in the labium, clitoris, or mons pubis. Most of the time a solitary mass is seen, but in 10% to 15% of cases multiple tumors are noted.² There are case reports of one patient with 26 lesions and another with 52 lesions.³ While the tumors may increase in size rapidly during pregnancy, they are generally slow-growing and usually take years to increase by 1 cm.¹ While most are asymptomatic, some symptoms may occur, including pain and pruritis.⁷

Differential Diagnosis

The differential diagnosis of granular cell tumors includes fibroma, sebaceous cyst, cancer, or xanthogranulomatous inflammation.² The tumor may be mistaken for squamous cell carcinoma, because more than half of cases show pseudoepitheliomatous hyperplasia.⁴

Microscopically the tumor consists of irregularly clustered groups of large epithelioid-appearing cells separated by hyalinized stroma.² The cytoplasm has a distinct granular appearance due to the accumulation of lysosomes. While most lesions are well circumscribed, about 50% show poorly defined margins.⁷ The tumor cells are immunoreactive for S-100 protein, myelin basic protein, and carcinoembryonic antigen. They stain with periodic acid-Schiff and Sudan black B and are resistant to diastase.³ In trichrome preparations, the tumors stain magenta.³

Treatment

Treatment is with wide local excision. There can be significant risk for blood loss due to the location of the lesions, as well as scarring

if there are irregular margins. Due to the rarity of granular cell tumors, having a pathologist with expertise in gynecology is preferred.

Recurrence

Recurrence rates are 2% to 8% in benign lesions with clear margins and 20% with positive margins.³ Malignant tumors are seen in 1% to 2% of cases and are very aggressive; they may not be diagnosed until regional or distant metastasis occurs.¹⁻³ They may show necrosis, nuclear pleomorphism, spindling, and increased mitotic activity, or they may appear morphologically benign. The malignant tumors do not respond to radiation or chemotherapy; treatment is surgical.³ The average age of malignant granular cell tumor patients is 50.

CONCLUSION

Once diagnosed with a granular cell tumor, the patient must be counseled to follow up regularly with physical exams. She should alert her clinician if any growth recurs at the excision site or if any nodular growth develops elsewhere on the body.

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