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## Research Letter

## Giant congenital melanocytic nevus presenting as a huge vulvar mass: A rare case

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## Dear Editor,

Genital pigmented lesions arise mainly on the vulva (labia majora, labia minora, and clitoris), although they may occur less often on the perineum, pubic region, and male genitalia (penis and scrotum) [1]. A giant congenital melanocytic nevus is an extremely rare lesion, occurring in one of 20,000 newborns [1]. They present in young people with sex predilection for females. They are usually small (<5 cm) and pose cosmetic problems only [2]. Rarely, they may become very large to cause disturbance in personal and sexual life. The authors present a case of a 20-year-old woman with a huge pigmented vulvar mass of the size of a watermelon since birth.

She was married for 4 months with disturbed sexual life. There was no pain, pruritus, fever, or ulceration. The family history was negative for any melanocytic lesions. Examination showed multiple black hyperpigmented lesions all over the body. There was no pallor, clubbing, cyanosis, edema, lymphadenopathy, or hepatosplenomegaly. Local examination revealed hyperpigmented areas involving the external genitalia, groin, and upper thigh (bathing-trunk/garment distribution; Figure 1A). There was a huge vulvar mass measuring 20 cm × 10 cm × 10 cm (Figure 1B). The pelvic examination was normal. Tests for filariasis, malaria antigen, leishmaniasis, syphilis, venous thrombosis, and human immunodeficiency virus were negative. Pap smear showed no intra-epithelial lesion. A biopsy of the mass showed an intradermal melanocytic nevus. Simple vulvectomy was performed under general anesthesia. The incision was made 2 cm from the base of the mass and resection done until a clean fat layer was visible. Proper reconstruction of the mons and vulva after excision of mass was performed (Figure 1C). The histopathological report revealed a

giant congenital nevus of the vulva. Multiple sections showed a compound nevus (intra-dermal with junctional activity) with evidence of deep tissue involvement consistent with a congenital nevus. No cytological atypia or mitosis was noted. The cells were immunopositive for S-100, melan-A, and HMB45 (Figure 2). After surgery, her sexual life changed dramatically and she conceived spontaneously after 8 months.

A melanocytic nevus is a variant of common nevi that exhibit distinct morphological features. It can be junctional, intra-dermal, or compound [2]. Our case had a compound nevus without cytological atypia. There is a 4–8% risk of developing malignant melanoma. These are usually small lesions ranging from 0.5 cm to 5 cm in size, but in our case, the lesion was very large measuring 20 cm × 15 cm × 10 cm. Congenital nevi show the presence of melanocytes around and within hair follicles, sweat and sebaceous glands, and their ducts. Melanocytes extend in between the collagen bundles and into the reticular dermis and subcutis. The nevus starts as a lentigo, which then turns into a pure junctional nevus with regular nests confined to the tips of rete pegs, to be followed by a compound phase, with finally the formation of a purely intra-dermal form, with senescence.

Friedman and Ackerman [3] first described an atypical melanocytic nevus of genital type in a series of seven unusual vulvar nevi. A previous study by Clark et al [4] has described clinical and histological features of genital lesions and recommended local excision as the preferred treatment.

Arao et al [5] reported a giant congenital nevus in a 32-year-old woman who had a massive pigmented tumor of the vulva. It grew over a period of 8 years and was histologically composed of benign nevus-like cells with focal areas of extensive fibrous response. Tumor cells were positive for S-100 protein and antihuman melanoma antibody (MoAb 225, 28S) stain.

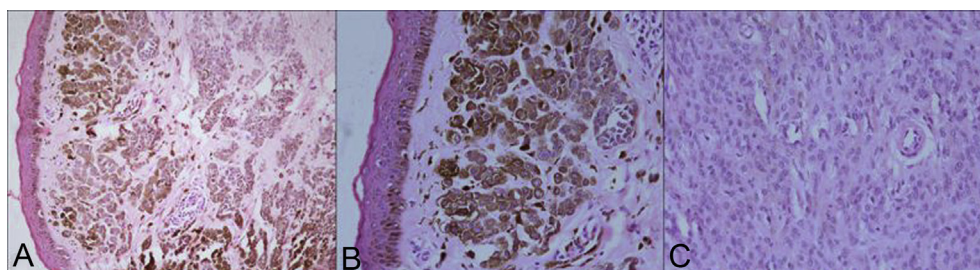
This case highlights the occurrence of a congenital melanocytic nevus as a large vulvar mass that hampered the sexual life of the patient. Ours is the second reported case of such a large congenital nevus. Biopsy remains the mainstay of diagnosis in cases of vulvar lesions along with the aid of immunohistochemistry. Conservative surgery in the form of local excision with reconstruction of external genitalia is the preferred treatment. These lesions have rare local

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**Figure 1.** (A) Multiple small hyperpigmented lesions all over the body and bathing trunk/garment distribution of pigmentation. (B) A huge vulvar mass (20 cm × 15 cm × 10 cm). (C) External genitalia after surgical reconstruction.



**Figure 2.** (A) The dermis shows nests of melanocytic cells; however, the epidermis and dermoepidermal junction are not involved (H&E, 100×). (B) Nests of round to oval nevus cells show heavy pigmentation in the superficial portion (H&E, 200×). (C) Cells in the deep portion show less pigmentation. No significant atypia or mitosis is evident (H&E, 200×). H&E = hematoxylin and eosin.

recurrence and no further adverse outcome as they have low malignant potential.

### Conflicts of interest

The authors have no conflicts of interest relevant to this article.

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