Immature teratoma of vulva in a 15-year-old girl

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Received: December 31, 2015  Accepted: February 10, 2016  Online Published: February 24, 2016

DOI: 10.5430/crcp.v3n1p74  URL: http://dx.doi.org/10.5430/crcp.v3n1p74

ABSTRACT

Background: Immature teratoma (IT) of vulva (V) has been reported only once.

Case report: A 15-year-old girl presented a vulvar tumor, and tumorectomy was performed. The tumor measured 4 cm × 4 cm × 5 cm and multiply cystic. The cysts contained mucinous fluid. The tumor was well demarcated from surrounding tissues and tumorectomy was easy. Microscopically, the tumor was multicystic neoplasm composed of mucin-bearing columnar mature epithelial cells, cellular immature mesenchymal zone beneath the columnar epithelial cells, immature cartilage, and mature connective tissue containing vasculatures. Immunohistochemically, the immature mesenchymal elements of the tumor were positive for p53, Ki67 (labeling = 30%) and vimentin, but negative for smooth muscle actin, myoglobin, HHF-35 caldesmon, and S100 element. No immature neural elements were seen. Since three germ elements were seen and some showed immature features, a diagnosis of IT was made.

Conclusions: A very rare case of IT of V is reported. This is the second case of IT in this location.

Key Words: Vulva, Immature teratoma, Pathology

1. INTRODUCTION

Teratoma is defined as a group of tumors composed of cells with more than two germ layers. Tumors consisted of more than two cell types of one germ layer is called mixed tumor. Teratoma occurs most commonly in genital organs i.e. ovary followed by testis. Teratoma occurs also in extra-gonadal sites where germ cells aberrantly migrate during development. Such extra-gonadal teratoma tends to develop in the midline of the body, such as brain and mediastinum. Teratomas are classified as mature (benign) and immature (malignant) teratoma (IT). The former consisted of mature cells without atypia, while the latter of immature cells particularly immature neuronal cells. Tumors of vagina are relatively rare, and most are condyloma acuminateum and other benign skin tumors. Malignant tumors of vulva (V) are mostly of epithelial origin. IT of V is extremely rare; only one case report of IT of V has been reported.

2. CASE REPORT

A 15-year-old girl presented a vulvar tumor, and tumorectomy was performed. The tumor measured 4 cm × 4 cm × 5 cm, and was soft and multiply cystic (see Figure 1). Grape-like projections (botryoid) were occasionally seen within the large cysts. The cysts contained mucinous fluid. The tumor was well demarcated from the surrounding tissues, and a tumorectomy was therefore easy.

Microscopically, the tumor was located under squamous epithelium of V (see Figure 2A). The tumor was a multicystic neoplasm composed of mucin-bearing columnar mature epithelial cells, cellular immature mesenchymal zone beneath the columnar epithelial cells, immature cartilage, and mature connective tissue containing vasculatures (see Figure 2B). The mature epithelial cells formed a layer of mucin-producing columnar epithelial cells. The larger cysts showed
intracystic grape-like projections (see Figure 2C). The immature mesenchyme formed a layer under the mature columnar cells, and simulated fetal developing mesenchyme\cite{8,9} or showed atypia such as hypercellularity and nuclear hyperchromasia (see Figure 2D). The cartilage was similar to developing fetal cartilage (see Figure 2E). No immature neural elements were seen.

An immunohistochemicaal study was done with the use of Envision method\cite{10}. The immature mesenchymal elements were positive for p53 (see Figure 2E), Ki67 (labeling = 30\%) (see Figure 2F) and vimentin, but negative for alpha-smooth muscle actin, myoglobin, HHF-35, caldesmon, and S100. Since three germ elements were seen and some showed immature histochemical and Immunohistochemical features, a diagnosis of IT was made. The patient is now free from tumors, and will be followed up strictly.

Figure 1. Gross features of tumor of vulva. The tumor is well defined and no invasive features are seen. The tumor is multicystic. Mucus is seen within the cysts. Grape-like protrusions (botryoid) are present in the cysts focally.

Figure 2. Histological (A-E) and Immunohistochemical (F, G) study of tumor of vulva
A: Low power view shows a cystic tumor (right) under squamous epithelium (left) of vulva. HE, ×20. B: The tumor is composed of the following three areas: 1) a layer of mucus-producing columnar mature epithelium, 2) subepithelial layer of immature mesenchymal, and 3) connective tissue with vasculatures. HE, ×40. C: The view shows intracystic grape-like protrusions of the immature mesenchymal element. HE, ×80. D: The immature mesenchymal element resembles fetal mesenchyme and shows atypical features such as hyperchromasia, hypercellularity and mitotic activity. HE, ×250. E: Cartilagenous element in the tumor is similar to fetal developing cartilage. HE, ×100. F, G: Immunohistochemistry. The immature mesenchymal element is positive for p53 protein (F) and Ki67 antigen (labeling index = 30\%) (G). F: ×150. G: ×100.
3. DISCUSSION

Although the author thinks the present case is IT of V, some other tumors such as Bartholin’s gland cystadenoma and sarcoma botryoides (embryonal rhabdomyosarcoma) can be possible. The present case is not the former. Also, the present case is not sarcoma botryoides because partly of negative muscular antigens.

Most of the immature elements of IT are neural. The present case showed no immature neural elements, but demonstrated immature mesenchymal and cartilaginous elements. The immaturity of both elements are reflected by high Ki67 labeling and positive p53 protein probably demonstrating p53 mutations.

4. CONCLUSION

A very rare case of IT of V is reported. This is the second case of IT in this location.

CONSENT

Informed consent was obtained from the patient. The publication was approved by the Ethical Committee of the Hospital.

CONFLICTS OF INTEREST DISCLOSURE

The author declares no conflicts of interest.

REFERENCES