Trichoblastic carcinosarcoma arising from the vagina: A case report with comprehensive immunophenotypic analysis

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Introduction

Carcinosarcomas are biphasic malignant neoplasms composed of epithelial (carcinomatous) and mesenchymal (sarcomatous) elements. To date, only five cases of trichoblastic carcinosarcoma have been reported, which all are of cutaneous primary. We present the first reported case of primary vaginal trichoblastic carcinosarcoma with an extensive immunostaining panel for pathological diagnosis, demonstration of tumor differentiation and delineation of its histogenesis.

Case report

A 54-year-old post-menopausal woman presented with abdominal pain and difficulty in voiding for two weeks. A large mass at the upper vaginal canal was found on physical examination. A biopsy was performed showing scattered spindle cells with variable low-to-moderate cellularity set in a loose myxoid background. No epithelial element was present in the initial biopsy. Subsequent radical hysterectomy and bilateral salpingo-oophorectomy with upper vaginectomy was performed. Histology showed a lobulated tumor with a biphasic impression at low power. A low cellularity pale cuff formed by spindle cells, as seen in the biopsy, surrounded darker cellular areas composed of small and hyperchromatic germinative cells. The spindle cells showed overgrowth and formed diffuse sheets originating from the organized lobules. The germinative cells were arranged in whorls resembling hair follicles with nests and strands of hyperchromatic epithelium extending from the follicles. The germinative components were surrounded by trichogenic stroma, evidenced by CD10 positivity, which merged imperceptibly with the spindle cell sarcomatous component. A detailed immunoprofile of each neoplastic element is listed in table 1. Both the spindle cell (sarcomatous) and germinative (carcinomatous) components were morphologically atypical, mitotically active and infiltrated the vaginal muscularis. All resection margins were clear. The patient remained disease free at 3 months' follow-up.

	Follicle	Trichogenic stroma	Germinative epithelium	Spindle cell
p16	+	+	weak	+
Bcl-2	+	+	+	weak
TLE1	+	+	-	+
Vimentin	+	+	-	+
CD56/NCAM	+	+	-	weak
TDAG51	+	+	-	-
EMA	weak	weak	-	-
CD10	-	+	-	-
p40	-	+	+	-
AE1/3	weak	-	+	-
CK5/6	-	-	+	-
34βE12	-	-	+	-
ER	-	-	-	-
PAX8	-	-	-	-

Table 1. Immunoprofile of neoplastic elements in trichoblastic carcinosarcoma

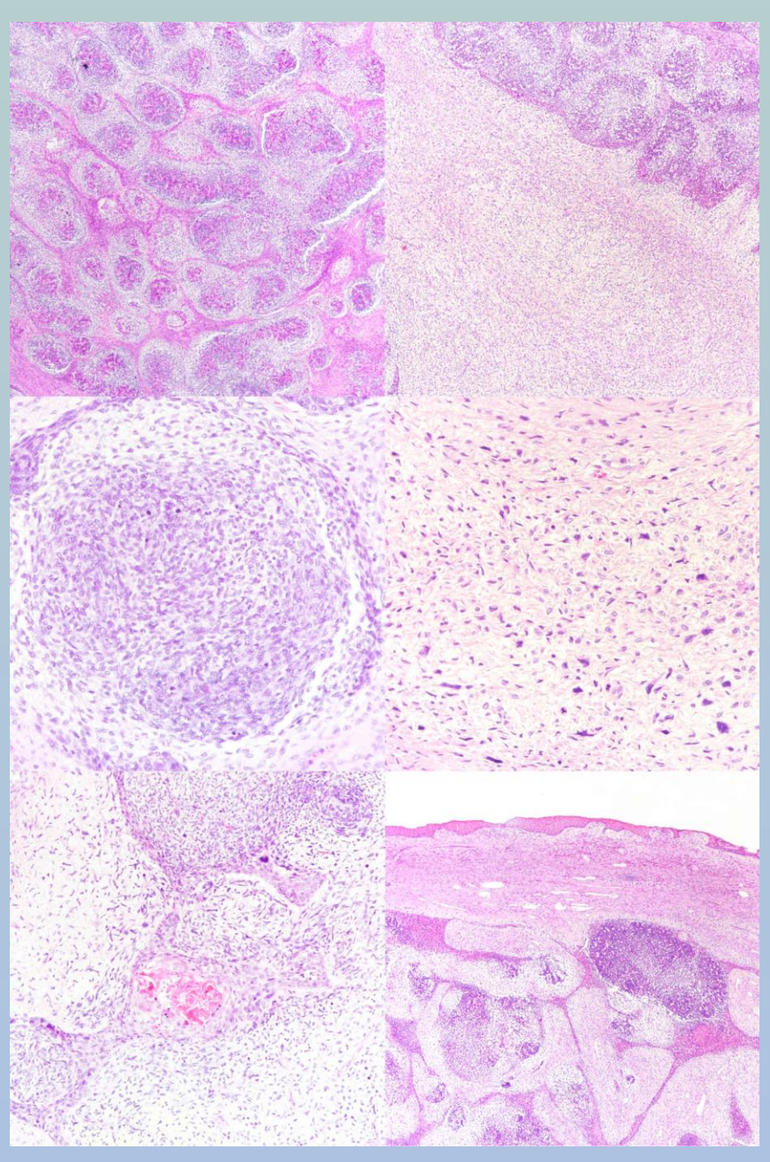

Figure 1. H&E sections of the tumor

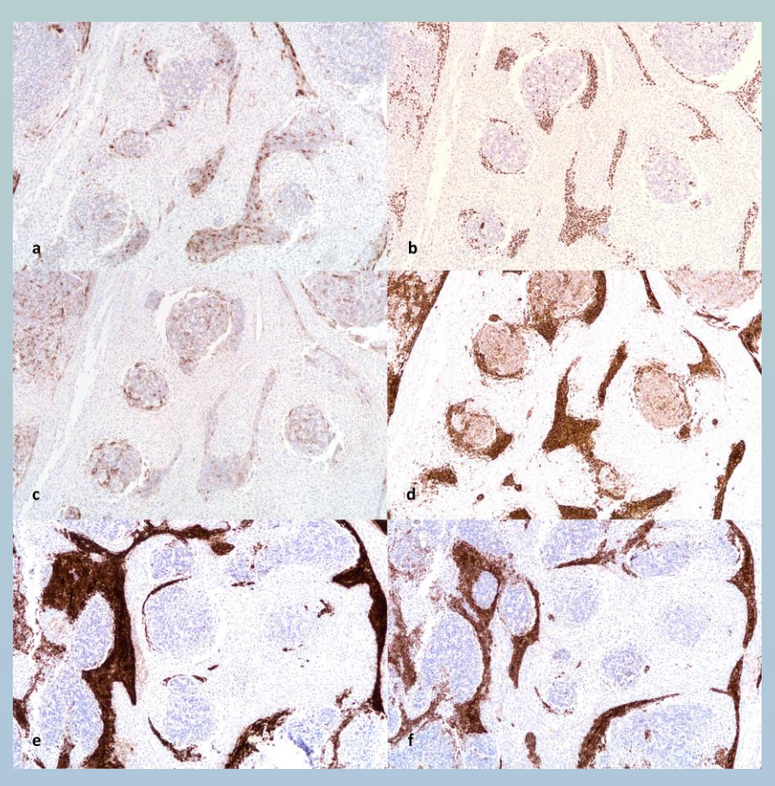

Figure 2. Immunostains demonstrating trichogenic stroma and epithelial differentiation

First row (left to right): a) CD10, 40x magnification, b) p40, 40x magnification; second row (left to right): c) EMA, 40x magnification, d) AE1/3, 40x magnification; third row (left to right): e) CK5/6, 40x magnification, f) 34βE12, 40x magnification

Discussion

An isolated case report of vulval adnexal tumor with extensive vaginal involvement has been reported in the literature. However, in our case, the tumor was located in the upper vagina without evidence of vulval/perineal skin connection on clinical examination, imaging or surgical exploration. Cutaneous adnexal tumors are also observed to develop from ovarian teratomas, but no ovarian pathology was seen in our case. As such, this case most likely represents a trichoblastic carcinosarcoma of vaginal origin. Presence of sebaceous glands and hair follicles have been reported in the uterine cervix and vagina, which were postulated to originate from congenital misplacement or represent acquired metaplastic changes. Primary mucosal skin adnexal tumors, of the gynecologic tract and other extra-cutaneous sites, are also well recognized in the literature. It is possible for the tumor in our case to have been arisen from ectopic adnexal structures.

Conclusion

Recognition of cutaneous-type carcinosarcoma is imperative as disease behavior and optimal treatment may be different from conventional gynecologic or visceral carcinosarcomas. Histological clues suggesting cutaneous differentiation include the lack of a high-grade serous or endometrioid carcinomatous component and evidence of epidermal or adnexal differentiation. Histologic and immunohistochemical evidence of trichogenic stroma, demonstrated by hair follicle markers (bcl-2, TLE1, CD56/NCAM and TDAG51), is important for establishing pathological diagnosis and ruling out vaginal epithelial and adnexal carcinomas.