Spermatic Cord Paraganglioma With Histologically Malignant Features

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Paragangliomas occur extremely rarely in the spermatic cord. A 40-year-old man presented with a scrotal mass that was diagnosed as spermatic cord paraganglioma with malignant histological features. To our knowledge, this is the first case of spermatic cord paraganglioma presented with malignant histological features evaluated by histological scoring. Careful evaluation of histological features and systematic evaluation should be considered for patients with spermatic cord paragangliomas. UROLOGY 93: e7–e8, 2016. © 2016 Elsevier Inc.

CASE PRESENTATION

A 40-year-old man presented with a painless mass in the left scrotum with no accompanying symptoms. Genital sonography revealed a mass in the spermatic cord (Fig. 1A). The mass was removed with unilateral orchiectomy. (Fig. 1B). The pathological specimen was diagnosed as extra-adrenal paraganglioma with histological features of malignancy (Fig. 1C-F). Whole body positron emission tomography–computed tomography of the patient did not reveal other lesions. Spermatic cord paragangliomas are extremely rare, with only 9 cases reported in the literature to date.[1-5] Compared with previously reported cases (Supplementary Table S1), the present case showed histological features of malignancy (Pheochromocytoma of the Adrenal gland Scaled Score [PASS] 6) with moderate differentiation (Grading system for Adrenal Pheochromocytoma and Paraganglioma [GAPP] score 4) (Table 1). Currently, there are no standardized histological criteria. However, scoring systems such as PASS[6] and GAPP[7] have been proposed to provide clarification. To evaluate the histological features systematically, we used PASS and GAPP scores for the present case. To our knowledge, this is the first case of spermatic cord paraganglioma with histological features of malignancy classified using histological scoring. The clear identifier of malignant paraganglioma is the presence of metastasis. At 1-year follow-up, the patient showed no evidence of recurrence or metastasis. Therefore, we report this tumor as an extra-adrenal paraganglioma with "histologically" malignant

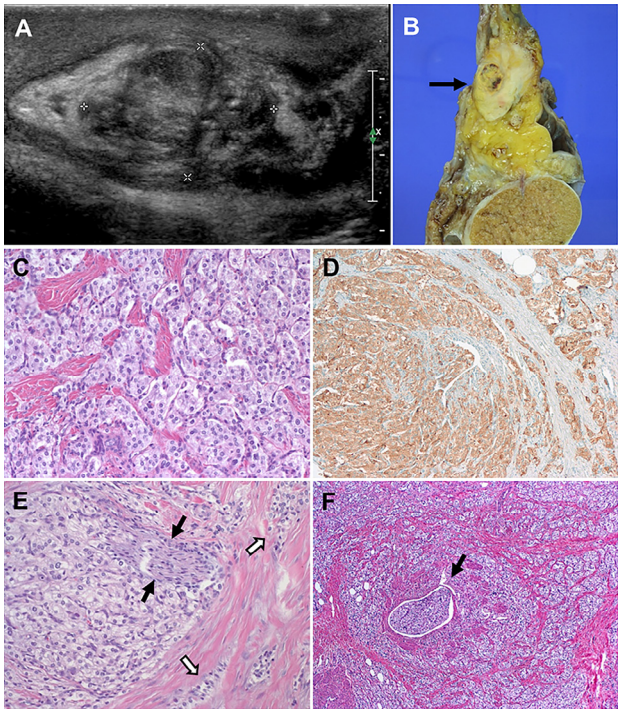

Figure 1. **(A)** Ultrasonographic imaging. Ultrasonography shows an ill-defined lobulated mass with heterogeneous echogenicity in the left spermatic cord. **(B)** Gross features. A well-defined, ovoid shaped, nonencapsulated, pale tan-colored firm mass measuring 1.8 × 1.3 cm with central hemorrhagic and ischemic necrotic foci in the spermatic cord is shown (arrow). **(C)** Histologic features. The tumor cells are arranged in well-defined nests ("Zellballen pattern") (H&E stain, 200×). **(D)** Immunohistochemical stain. The tumor cells showing positive reaction for synaptophysin (100×). The cells showing positive reactivity for chromogranin (not shown); the sustentacular cells are positive for S-100 (not shown). **(E)** Perineural invasion. Perineural invasion of tumor is shown with black arrows (H&E stain, 200×). Focal tumor cell spindling is also seen (white arrows). **(F)** Vascular invasion. Note the vascular invasion of tumor present in the medium-sized vein (black arrow, H&E stain, 100×).

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Table 1. PASS and GAPP score of the present case

PASS parameters*		GAPP parameters†	
Large nests or diffuse growth	0/2	Histologic pattern: Zellballen	0/2
Central or confluent tumor necrosis	0/2	Cellularity: moderate	1/2
High cellularity	0/2	Comedo necrosis: absent	0/2
Cellular monotomy	2/2	Vascular or capsular invasion: present	1/1
Tumor cell spindling	2/2	Ki67 labeling index: 3-5%	2/2
Mitotic figures 0-1/10 HPF‡	0/2	Catecholamine type: nonfunctioning type	0/1
Atypical mitotic figure	0/2		
Extension into adipose tissue	1/1		
Vascular invasion	1/1		
Capsular invasion	0/1		
Profound nuclear pleomorphism	0/1		
Nuclear hyperchromasia	0/1		
Total score	**6/20**	**Total score**	**4/10**

GAPP, Grading system for Adrenal Pheochromocytoma and Paraganglioma; HPF, high-power field; PASS, Pheochromocytoma of the Adrenal gland Scoring Scale.

* ≥4, potentially malignant biological behavior; <4, benign biological behavior.

† 0-2: well-differentiated type; 3-6: moderately differentiated type; 7-10: poorly differentiated type.

‡ ×400.

features, avoiding the term "malignant" paraganglioma. We advise that patients should be carefully assessed when evaluating spermatic cord paragangliomas.

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APPENDIX

Supplementary Data

Supplementary data associated with this article can be found, in the online version, at http://dx.doi.org/10.1016/j.urology.2016.03.014.