A Long-Standing Primary Vaginal Paraganglioma—Coexisting with Esophageal Carcinoma

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Sir,

Paraganglioma is a rare neoplasm derived from the neural crest cells residing in the paraganglia of the autonomic nervous system. Primary vaginal paraganglioma is extremely rare. [1]. Herein, we present a unique case of a vaginal paraganglioma which was radiologically interpreted as a metastatic nodule in a young female, which is a known case of esophageal carcinoma.

A 28-year-old married female presented with dysphagia to solid foods was diagnosed as having keratinizing squamous cell carcinoma of the lower esophagus on biopsy 5 years back. She received neoadjuvant chemoradiotherapy (NACRT). Post-NACRT PET-CT revealed moderate to intense FDG (fludeoxyglucose) uptake with decline in SUV max to 6.7 indicating a favorable response to chemotherapy. However, PET-CT revealed an intense FDG-avid (SUV max = 20.9) homogeneous soft tissue lesion measuring 2.1×2.1 cm in the left lateral vaginal wall with well-maintained fat planes with the adjacent urinary bladder wall (Fig. 1a). On per-vaginum examination, a firm, well-defined mass measuring 2×3 cm was noted along the middle one-third of the left lateral vaginal wall, though she did not have any gynecological complaints. Fine-needle aspiration cytology (FNAC)

from the vaginal mass did not reveal any malignant cells, and the cells were interpreted as histiocytes. Following this, she underwent trans-hiatal esophagectomy which confirmed the diagnosis of keratinizing squamous cell carcinoma.

Her follow-up PET-CT scan post-surgery showed similar FDG uptake in the vaginal mass, without any increase in its size. However, the most recent PET-CT scan showed increase in the FDG uptake. Contrast-enhanced computed tomography (CECT) showed the vaginal lesion to be heterogeneously enhancing soft tissue mass measuring 3.6×2 cm with increased vascularity and a clinical possibility of metastatic tumor deposit was favored. USG-guided FNAC from the mass revealed predominantly blood along with few histiocytic collections (Fig. 1b). Finally, the mass was surgically excised and the vaginal wall was reconstructed. Grossly, it measured 3×3 cm and had a firm and brown-colored cut-surface. On microscopic examination, the tumor exhibited a classical Zellballen pattern comprising of nests and islands of tumor cells with a prominent intervening fibrovascular stroma (Fig. 1c). The tumor cells were large and polygonal shaped with fine-stippled chromatin, inconspicuous nucleoli, and abundant granular eosinophilic to clear cytoplasm with well-defined cell membranes. Tumor cell nests were bordered by few spindle-shaped sustentacular cells (Fig. 1a). At the periphery, the blood vessels were even more prominent and hyalinized. No mitosis or necrosis was noted. On IHC, the tumor cells were strongly positive for chromogranin (Fig. 1d), synaptophysin (Fig. 1e), neuron-specific enolase (NSE), and CD 56. S-100 protein highlighted the sustentacular cells (Fig. 1f), and pan-cytokeratin was negative. Based on these morphological features, a diagnosis of primary vaginal paraganglioma was made. Retrospective review of FNAC smears revealed few singly scattered tumor cells with round nuclei and abundant granular cytoplasm consistent with

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Fig. 1 **a** PET-CT scan showing an intense FDG-avid soft tissue lesion along the left lateral vaginal wall (arrow). **b** Cytology smear (May-Grünwald Giemsa $\times 400$) showing large polygonal cells with round to oval nuclei and abundant granular cytoplasm. Microphotograph (H&E $\times 100$) showing nests of tumor cells separated by delicate fibrovascular

stroma. **c** Tumor cells are large polygonal with abundant eosinophilic granular cytoplasm with few sustentacular spindle cells (arrow). IHC showing cytoplasmic chromogranin (**d**) and synaptophysin (**e**). **f** S-100 protein highlights sustentacular cells

paraganglioma (Fig. 1b). Currently, the patient is disease free and is on close follow-up.

Paraganglioma, a benign neuroendocrine tumor, rarely involving the female genital tract, is confined to few case reports in the uterus, ovary, vulva, vagina, and broad ligament [1]. In literature, only eight cases of vaginal paraganglioma have been reported till date [1–3].

Depending upon their function to synthesize, store, and secrete catecholamines, neurohormones, and peptides, the paragangliomas are divided into functional and non-functional subtypes [1]. The index case was a non-functional paraganglioma and fortunately, no hypertensive crisis was observed even on repeated FNAC.

The pre-operative diagnosis is highly challenging particularly when it is asymptomatic/non-functional. Clinically, the differential diagnosis especially in young females includes common vaginal neoplasms like leiomyoma, embryonal rhabdomyosarcoma, and hemangioma [1]. CECT reveals a contrast-enhancing hypervascular soft tissue mass arising from the vagina [4]. PET-CT can aid in identifying the malignant or metastatic disease. Blood and urine catecholamine and nor-adrenaline levels in the clinical setting of hypertension can also aid in the diagnosis of a functional paraganglioma.

The differential diagnoses in the index case were metastatic esophageal squamous cell carcinoma. A recent increase in FDG uptake in the mass led to the clinical suspicion of metastasis. The tumor cells were completely negative for cytokeratin and diffusely expressed neuroendocrine markers, ruling out the possibility of metastatic squamous cell carcinoma.

These tumors require a close follow-up as around 15–50% of these tumors at other locations have shown malignant behavior [5]. Irrespective of histopathology, the malignant transformation is indicated by recurrence, metastasis to the lymph nodes and/or non-chromaffin tissues, or invasion of the adjacent organs.

We conclude that vaginal paraganglioma is an extremely rare tumor which can mimic other neoplasms. So, histopathological and immunohistochemical examination along with periodic and close follow-up are mandatory in such cases.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

Financial Disclosure None

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