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Primary small cell neuroendocrine carcinoma of vagina - A rare entity at a rare site

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ABSTRACT

Primary small cell neuroendocrine carcinoma of the vagina is extremely rare and its clinical behavior is aggressive. To our knowledge, only 25 patients with this tumor have been reported in the literature to date .We describe a case of primary neuroendocrine carcinoma arising from the anterior vaginal wall of a 60 year old woman who presented with dysuria and growth in vagina. The mass was 2 x 1 x 1 cm in size. Histopathological study revealed stratified squamous epithelium with underlying tumor tissue arranged in solid sheets. The cells were small sized with scanty ill defined cytoplasm, hyperchromatic nuclei with coarse to finely granular chromatin and inconspicuous nucleoli. The typical neuroendocrine architecture was absent excluding the possibility of typical and atypical carcinoid. Immunohistochemistry for neuroendocrine marker confirmed the diagnosis. Primary small cell neuroendocrine carcinoma of vagina is very rare. Infrequently small cell component is seen in association with a primary vaginal adenocarcinoma. In the present case there was no associated adenocarcinomatous component in the vagina. Chest X-ray and lower genital tract of the patient also did not reveal any abnormality. So a confirmed dignosis of primary small cell neuroendocrine carcinoma of vagina was rendered.

INTRODUCTION

Primary small cell neuroendocrine carcinoma of the female genital tract is a rare entity which has been reported to originate in the cervix, endometrium, ovary , vagina and vulva in decreasing frequency. Primary small cell neuroendocrine carcinoma of the vagina is extremely rare and its clinical behavior is aggressive and carries a very poor prognosis, even with the current therapeutic modalities. The histologic, immunohistochemical and ultrastructural profiles are similar to small cell carcinoma originating in other primary sites. To our knowledge, only 25 patients with this tumor have been reported in the literature to date [1].

CASE HISTORY

A 60 years old female para 4 with 8 months history of difficulty in voiding urine was found to have a discoid vaginal mass in the anterior vaginal wall in close proximity to the introitus without any cervical involvement. Her pervaginum revealed no abnormality.

The patient had no significant medical or surgical past history. There was no history of smoking, alcohol intake and drug abuse. Her human immunodificiency virus status was negative. Bone scan and chest X-ray were negative for any mass.Her pap smear was inflammatory without abnormal cells. The mass was clinically diagnosed as a vaginal polyp and was excised under local anesthesia.

The gross specimen received was greyish white discoid mass measuring 2x1x1cms. Cut surface was solid with small hemorrhagic areas(Fig 1). Histopathology section showed stratified squamous epithelium with underlying tumor tissue arranged in nests and cords of closely packed small cells with scanty eosinophilic cytoplasm and indistinct cell border, centrally placed nuclei with fine granular chromatin and inconscpicuous nucleoli(Fig 2).

Absence of typical neuronendocrine architecture excluded the

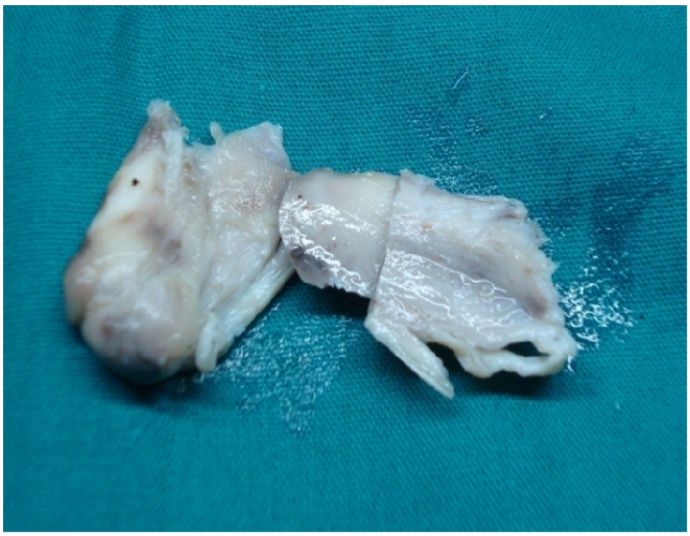

Fig 1 : Gross photograph of mass removed from vagina

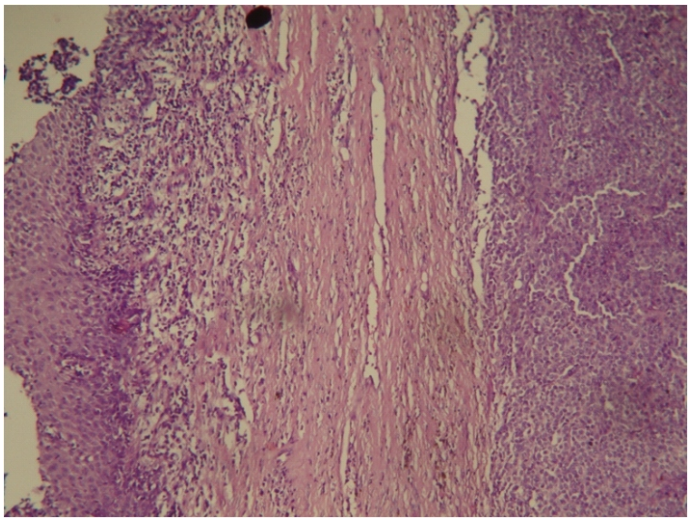

Fig 2 : Histopathology showing stratified squamous epithelium with underlying small dark tumor cells. H&E stainx100x

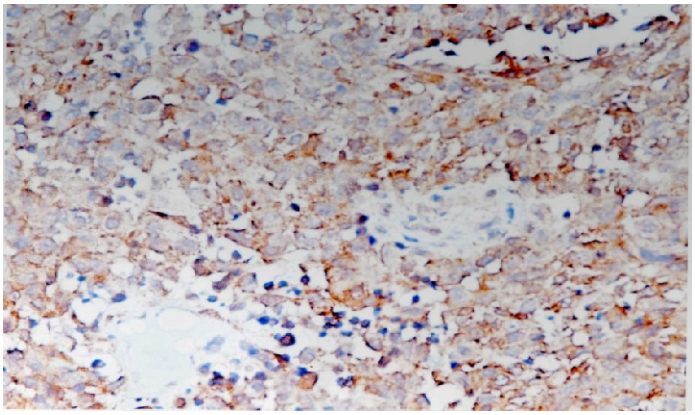

Fig 3 : Photomicrograph showing Immunohistochemistry positivity for neurone specific enolase (NSE)

possibility of typical and atypical carcinoid. Immuno-histochemical staining for neuron specific enolase showed 2 + immunoreactive score positivity in tumour cells (Fig 3) .

DISCUSSION

Primary small cell neuroendocrine carcinoma of the vagina is a rare and aggressive neoplasm first reported in 1984 by Scully et al . The histologic findings are those of typical and classic small cell carcinoma, consisting of tightly packed, small round and oval cells with scanty cytoplasm, ill defined cell borders, fine granular nuclear chromatin and absent or inconspicuous nucleoli. Nuclear moulding was observed in few foci with no definitive differentiation into epithelial structure.

Small cell carcinoma of the vagina must be distinguished from the small cell variant of squamous cell carcinoma and basaloid carcinoma. The small cell variant of squamous cell carcinoma is a poorly differentiated carcinoma with small tumor cells that retain morphologic characteristics of a non-small cell carcinoma and shows focal squamous differentiation. This also must be distinguished from neoplasms in which there is a mixture of squamous cell carcinoma and true small cell carcinoma. The small cell variant of squamous cell carcinoma lacks the characteristic nuclear features of small cell carcinoma. The tumor cells show nuclei with coarse or vesicular chromatin, prominent nucleoli and more district cytoplasmic outline. Focal intercellular bridges may be seen.

The basaloid variant of squamous cell carcinoma and basaloid carcinoma are composed of tumor nests with prominent peripheral palisading, small monomorphic cuboidal to fusiform cells with hyperchromatic nuclei, fine granular chromatin, absent or only focal nucleoli, scant cytoplasm and high mitotic rate. In basaloid carcinoma neither intercellular bridge nor individual cell keratinisation is present.

Immunohistochemical stain for neuroendocrine marks are usually negative. Accurate diagnosis of small cell neuroendocrine carcinoma and its distinction from non-small cell carcinoma will determine the patients management and prognosis.

The diagnosis of primary small cell carcinoma of the vagina is established after exclusion of a direct extension of the tumor from the cervix and metastasis from the lung and other extrapulmonary sites. Absence of any other mass in the cervix and negative X-ray of chest rules out the possibility

No consensus has been reached regarding its optimal therapy, since the disease is so rare and the current therapies have usually resulted in poor outcomes. The initial treatment options are usually based on the patients' clinical stage. For lesions that are small without evidence of metastasis, surgical resection with a partial or total vaginovulvectomy with bilateral inguinal and pelvic lymphadenectomy may be indicated, followed by chemoradiation[2]. Our patient with a small sized swelling belonging to stage 1 (confined to vagina) had received 5 cycles of chemotherapy comprising of cyclophosphamide, adriamycin and vincristine following surgical resection and is doing well. Given the limited clinical experience with this rare tumor, the therapeutic modalities with these patients should be explored.

CONCLUSION

Small cell carcinomas are extremely rare in extrapulmonary sites accounting for 1-2% of gynecological malignancies.[3,4] The histological, ultrastructural and immunohictochemical features are similar in both pulmonary and extrapulmonary sites.

The clinical course is aggressive and treatment is by surgery followed by chemoradiation.

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