Letter to Editor

An unusual case of aggressive small-cell neuroendocrine carcinoma of cervix in a young woman

Sir,

Neuroendocrine tumors comprise a broad family of tumors that arise from the diffuse neuroendocrine cell system. They usually arise in the lungs, bronchi, small intestine, appendix, rectum, pancreas, and thymus. The uterine cervix is a very unusual site accounting for 1.4% of all cervical cancers. These are highly aggressive tumors, characterized by early distant metastasis and worse prognosis compared to other histological types occurring in the cervix. Distant sites of recurrence, including lung and bone, are more common (28%) than local recurrence (13%).[1] Neuroendocrine carcinoma of cervix (NECC) is graded into three categories based on the morphology of tumor cells, mitotic index, and Ki-67 index. They are typical carcinoid (Grade I), atypical carcinoid (Grade II), and poorly differentiated carcinoma (Grade III). Poorly differentiated neuroendocrine carcinoma includes small-cell and large-cell neuroendocrine carcinomas.[1] Most NECCs of the cervix are small-cell carcinomas. We report a case of small cell neuroendocrine carcinoma of the cervix in a young woman diagnosed by histopathological examination because of its rare incidence and diagnostic challenge. A 32 year multiparous woman with regular 6/30 day menstrual cycles presented with intermittent bleeding per vaginum for 6 months. On examination, her vitals were stable; general, systemic, and abdominal examinations were normal. Per speculum examination revealed a polypoidal mass of 2 cm × 1 cm arising from the posterior lip of the cervix. Pap smear was negative for malignancy. Excision biopsy was performed, and histopathological examination of the excised mass revealed normal stratified squamous cervical epithelium with subepithelial stroma showing nests of small tumor cells with indistinct cell borders, scant cytoplasm, pleomorphic hyperchromatic nucleus, and indistinct nucleoli [Figures 1 and 2]. Immunohistochemical analysis showed positivity for synaptophysin and chromogranin A in small tumor cells which are consistent with small-cell neuroendocrine carcinoma. Our patient had early-stage disease as per FIGO staging and underwent radical hysterectomy followed by adjuvant chemotherapy with cisplatin and etoposide regimen. She is on regular follow up for the past 6 months without evidence of recurrence or metastasis. Histopathological examination and immunohistochemical analysis aid in accurate diagnosis of NECC. The median age of diagnosis is in the fifth decade of life. Small-cell neuroendocrine carcinoma of the cervix (SCNECC) exhibits clinical and biological characteristics of both cervical neoplasm (such as local aggressiveness and involvement of papillomavirus) and neuroendocrine small-cell cancer of any site (such as early dissemination and loss of heterozygosity at different loci). The FIGO stage of the tumor is the most important prognostic factor associated with survival.[1,2] Our case

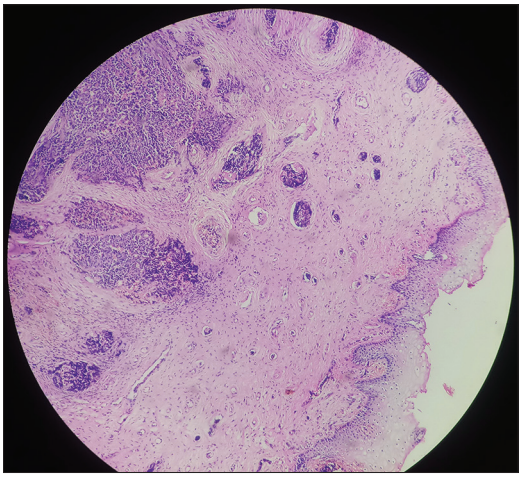

Figure 1: Microphotograph showing stratified squamous epithelium of cervix with subepithelial stroma showing nests of small tumour cells (H and E stain, ×40)

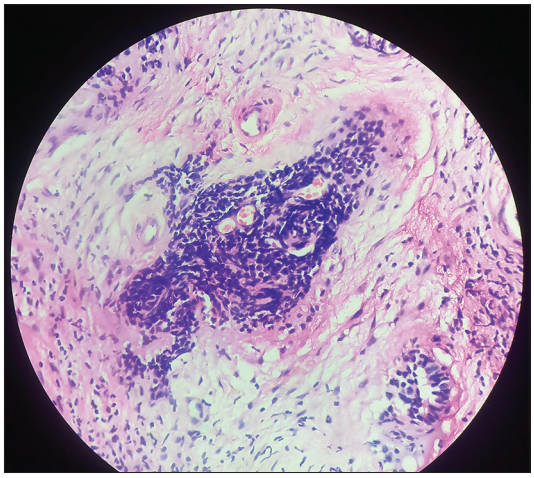

Figure 2: Microphotograph showing nests of small tumour cells with scant cytoplasm, pleomorphic hyperchromatic nuclei with indistinct nucleoli (H and E stain, ×400)

had a clinical presentation similar to the case reported by Saini *et al*.[3] SCNECC during pregnancy has also been reported.[2] As this tumor is rare, there is no standardized treatment protocol. These tumors are usually treated with radical hysterectomy and lymphadenectomy followed by adjuvant cisplatin plus etoposide chemotherapeutic regimen with or without radiotherapy.[2,4] As SCNECC is rare and highly aggressive with high rate of distant metastatic spread, it is important to recognize this histopathological entity, followed by detailed systemic evaluation and a multimodality therapeutic approach to maximize the survival.

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Conflicts of interest

There are no conflicts of interest.

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