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Cystic Endometrial Stromal Nodule with an Unusual Gross Morphology

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Case Report

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Abstract: Endometrial Stromal Tumours are rare mesenchymal neoplasms of the uterus, the cells of which resembles stromal cells of proliferative endometrium. Grossly, most common appearance of Endometrial Stromal Nodule is a circumscribed solid mass. We report a case of Endometrial Stromal Nodule which has undergone complete cystic degeneration and has acquired a unilocular tender coconut-like cystic appearance which is very unusual.

Keywords: Cystic, Endometrial stromal nodule, Endometrial stromal sarcoma, Gross morphology, Unilocular.

INTRODUCTION

Endometrial stromal tumours (ESTs) are rare mesenchymal neoplasms of the uterus. In 2014 WHO classification ESTs are subdivided into 4 groups- Endometrial stromal nodule (ESN), Low-grade endometrial stromal sarcoma(LGESS), High-grade endometrial stromal sarcoma (HGESS) and undifferentiated uterine sarcoma (UUS) [1]. The high grade ESS had been removed from the 3rd edition (2003) WHO classification scheme but has been reintroduced in the 4th edition.

WHO defines ESN as a benign endometrial stromal tumour that has a well circumscribed margin and is composed of cells that resemble proliferative phase endometrial stromal cells. Finger-like projections or immediately adjacent nests of tumour cells (measuring <3mm in greatest extent from the main mass and <3 in number are acceptable. Lymphovascular invasion excludes the diagnosis [1].

LGESS has permeative growth into the myometrium as tongue like projections and lymphovascular invasion. HGESS show destructive infiltration into the myometrium and shows >10 mitosis/10HPF, necrosis and pleomorphism but, there will be a focal area resembling LGESS. If there is no differentiation at all, the tumour is categorized in the UUS type.

Tumours are commonly submucosal, but may be intramural with or without connection to the endometrial cavity. Cyst formation may occur, which may be uni or multi-loculated but, predominantly cystic tumours are rare. We are reporting a case of ESN which presented with a unilocular cystic appearance because of complete cystic degeneration of the tumour.

CASE REPORT

49 year old lady admitted with complaints of lower abdominal pain and menorrhagia since last 6 months. Ultrasonogram of the abdomen showed bulky uterus with cystic areas in the uterus. Possibility of a fibroid with cystic degeneration was considered. Total Hysterectomy was done.

We received the total hysterectomy specimen in our department. Uterus was enlarged, more towards one side and measured 13X10.5X8 centimetres. (Figure 1a) On cutting open, the myometrium showed a well circumscribed unilocular cystic lesion measuring 6.5x5.5x6.5cms containing thin yellow serous fluid. The inner wall had a tender coconut-like appearance with a scrapable coating of pale yellow material measuring 2mms in thickness (Figure 1b).

Fig-1a: Gross appearance of Total hysterectomy specimen.

Fig-1b: Cut section of uterus showing the cystic cavity with tan yellow coloured inner wall.

No solid areas or papillary projections noted. There was no connection with the endometrial cavity. Endometrial canal measured 3cms with an endometrial thickness ranging 0.2-0.5 cm. An attached stump of fallopian tube measuring 1cm was also visualised on one side.

Grossly the differential diagnoses were leiomyoma with cystic degeneration and cystically dilated adenomyosis focus. The gross appearance also had a resemblance to a hydatid cyst, though it is very rare. Extensive sampling was done from the cyst wall and adjacent myometrium. Other areas were sampled as per routine.

Microscopic examination of the cyst wall showed a well circumscribed neoplasm with extensive cystic degeneration surrounded by myometrial smooth muscle. Apposed to the myometrial fibres, was a few layers of foam cell collection admixed with tumour cells. Beneath this, was several layers of tumour cells (Figure 2a, b, c). Many spiral arteriole-like vessels were

seen scattered among the tumour cells with whorling of the cells around it (Figure 2d). The tumour cells are composed of small round to oval/spindle cells with scanty cytoplasm and round or oval regular nuclei, in areas admixed with foam cells with multivacuolated cytoplasm. There was no cellular or nuclear pleomorphism. Mitosis was scanty (Figure 3a &b). Focal areas of intercellular oedema were present. Tumour was not infiltrating into the adjacent myometrium. No lymphovascular invasion noted. Tumour was not seen in any other area of the uterus. Other areas of myometrium showed foci of adenomyosis. Attached fallopian tube stump was histologically unremarkable.

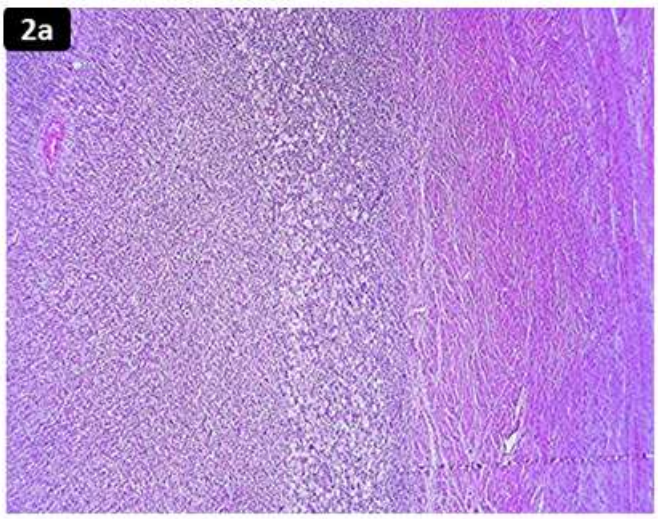

Fig-2a: Scanner view showing layering of myometrium, foam cell collection & tumour cells.
Note the absence of infiltration into the adjacent myometrium (H&E x40).

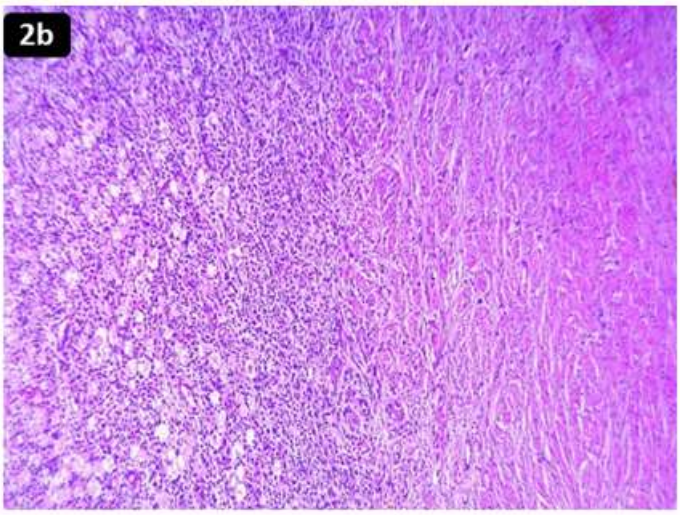

Fig-2b: Myometrial- tumour junction. Foam cells are seen admixed with tumour cells (H&E x100).

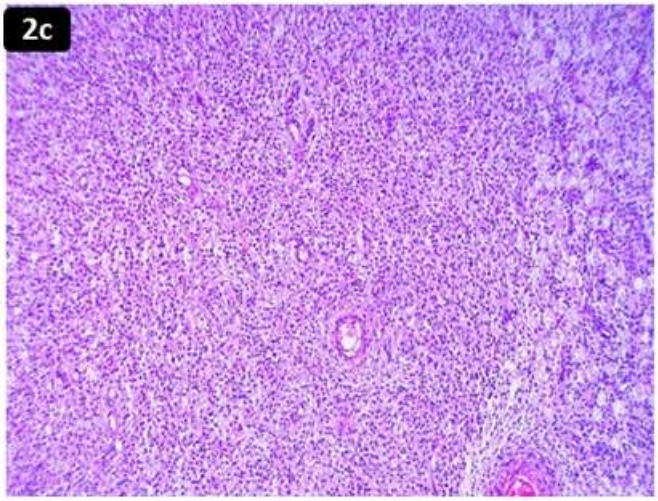

Fig-2c: Tumour cell layer with foam cell layer (H&E x100).

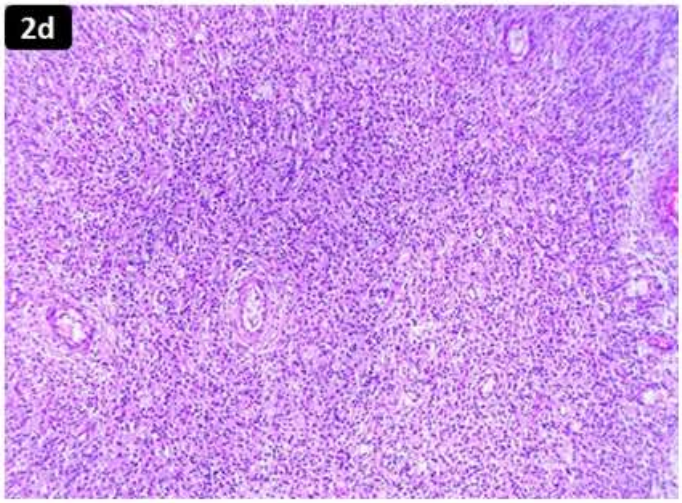

Fig-2d: Spiral arteriole like vessels within the tumour (H&E x100).

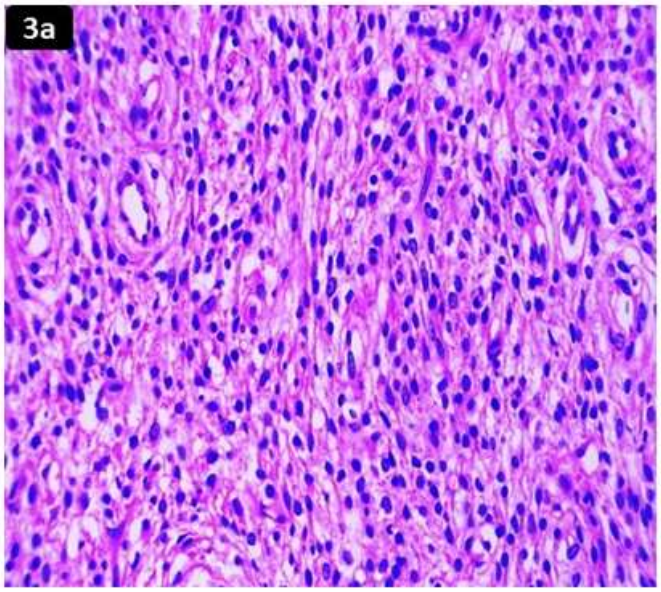

Fig-3a: Tumour cells showing the typical morphology (H&E x400).

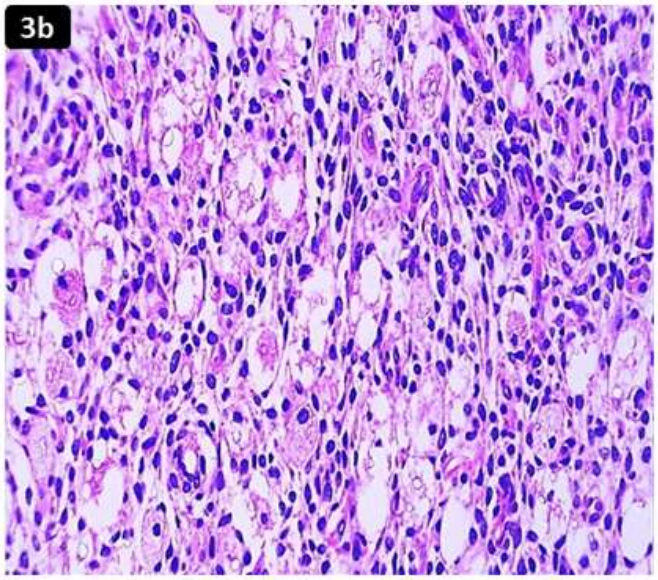

Fig-3b: Foam cells with multivacuolated cytoplasm and central nucleus, within the tumour (H&E x400).

Immunohistochemistry study was done for CD10 which showed strong diffuse positivity in tumour cells (Figure 4a&b). Markers for SMA (Figure 4c) & Desmin were negative in tumour cells. Ki 67 showed low proliferative index. CD 68 marker highlighted the scattered foam cells within the tumour (Figure 4d).

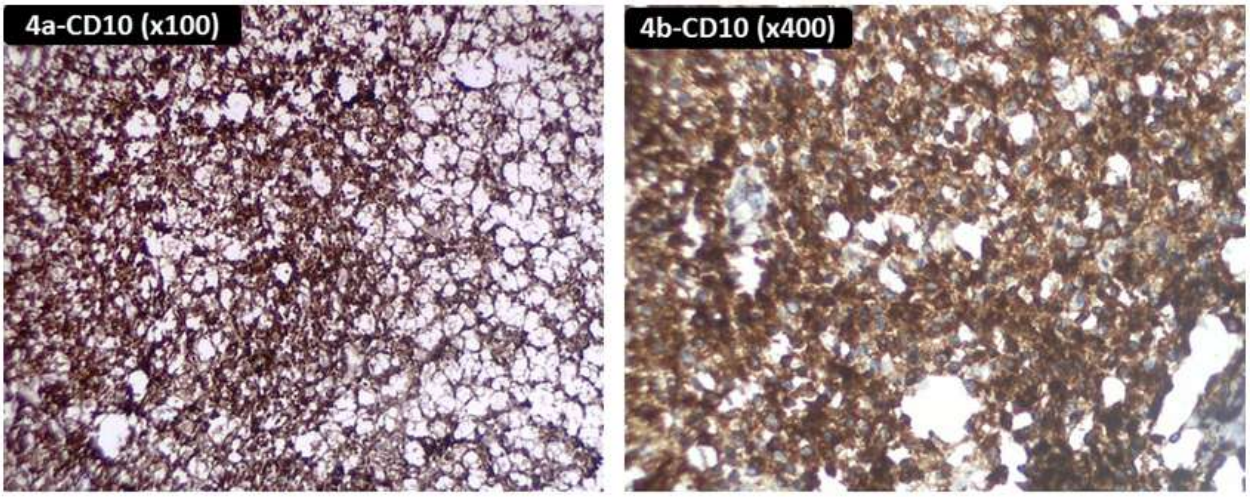

Fig-4a & 4b: IHC marker for CD10 (x100 & x400) showing strong diffuse positivity.

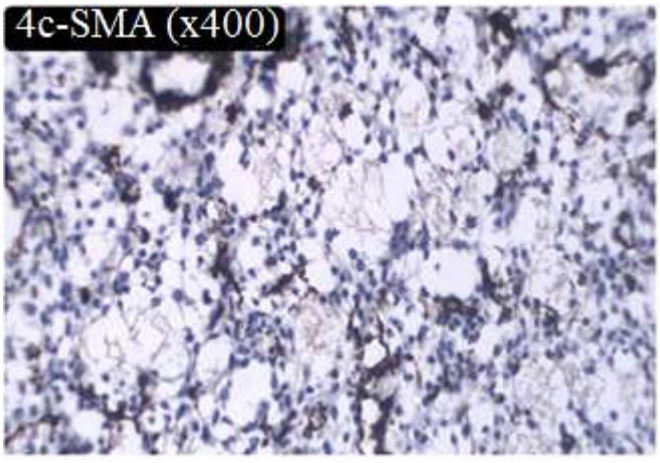

Fig-4c: IHC marker for SMA showing negative tumour cells (x400).

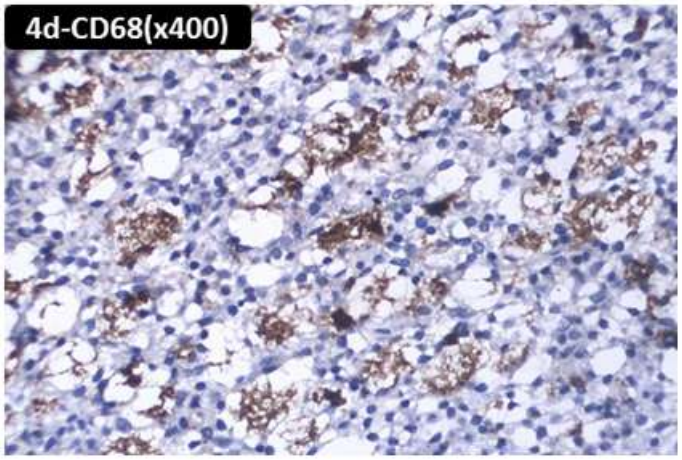

Fig-4d: IHC marker for CD68 showing scattered positive foam cells (x400).

DISCUSSION

ESTs occur in perimenopausal women. In a case series study by Dionigi A *et al*., ESN occurs over a wide age range, from 31 to 86 years with a mean of 53 years [2]. Our case was that of a 46 year old woman. But, it has been reported in younger age also [3, 4].

ESN is a well-circumscribed non-encapsulated, neoplasm, whereas low grade ESS is infiltrative with lymphatic vessel permeation. Finger-like projections or immediately adjacent nests of tumour cells measuring < 3 mm in greatest extent from the main mass and < 3 in number are acceptable. Lympho-vascular invasion excludes the diagnosis [1]. HG ESS and UUS show a destructive infiltrative growth into the myometrium. On rare occasions, low-grade ESS may appear deceptively well circumscribed on gross examination [5].

Completely cystic appearance of endometrial stromal nodule is very rare. Cases have been reported with small cystic spaces or partial cystic degeneration in ESN [6]. Anna Somma *et al*., [7] had described 2 cases and Delia Perez Montiel [4] 1 case of ESS with a multiloculated cystic appearance.

Cellular and highly cellular leiomyomas are differential diagnoses for a cystic tumour in the myometrium. These can also show cystic degeneration and is soft, yellow or tan yellow. In a study of cellular benign mesenchymal tumours of uterus by Olivia E *et al.*, [8] only one out of 6 tumours showed predominantly cystic pattern. Others were predominantly solid or solid and cystic. Microscopically also, HCL may mimic an EST. Presence of large thick muscular-walled blood vessels throughout the tumour is characteristic of highly cellular leiomyoma whereas in EST small spiral arteriole like vessels are seen. In EST, large thick blood vessels if present are typically seen close to the tumour-myometrium interface in nodules or close to largest masses of tumour in low-grade sarcomas probably representing entrapped pre-existent vessels [9].

In the present case, a few layers of foam cell collection admixed with tumour cells were noted which was highlighted by immunomarker for CD68. In the study of Oliva E *et al*., [8] also foam cell collection was noted and demonstrated by CD68 in the tumours. Foam cell collection is common in ESNs.

Various types of metaplastic change can occur in ESTs namely smooth muscle, clear cell, rhabdoid, fatty, and others, of which smooth muscle differentiation is the commonest. Differentiation towards Sex cord, endometrial gland etc can rarely occur. In our case metaplastic elements were not seen.

Immunohistochemical study shows strong diffuse positivity for CD10 marker in ESTs. But, diagnosis could not be entirely dependent on immunohistochemistry because smooth muscle tumours and PECOMAs can be positive for CD10 and ESNs can be positive for smooth muscle markers. But, usually ESNs will not be positive for more than one smooth muscle marker. Hence, even though CD10 is not a specific marker for EST, SMA & Desmin negativity with CD10 positivity rules out a highly cellular leiomyoma.

A definitive diagnosis of ESN can be rendered only after careful sampling and examination of the tumour border, which is only possible in a hysterectomy. Curetting specimens are not adequate for a definite diagnosis. Therefore, the final interpretation should be with correlation of the H&E morphology of the tumour and diagnostic evaluation is complete only in a total hysterectomy specimens [10]. Hence hysterectomy is the ideal modality of first- line management for correct diagnosis and planning of further treatment in peri/post-menopausal women. Conservative fertility-sparing management is advised for young patients.

CONCLUSION

Grossly, complete cystic degeneration of ESN is rarely encountered. Especially an appearance like a hydatid cyst is very rare. Hence any cystic lesion in the uterus should be histologically assessed and immunohistochemical studies should be done to confirm the diagnosis and exclude the mimicker lesions. Moreover, extensive sapmling of adjacent myometrium should be done to exclude a LGESS because, ESNs do not relapse but low-grade ESSs which have a low malignant potential are characterised by multiple recurrences even after many years. Metastasis has also been reported in LGESS. Hence it is important to differentiate the two for therapeutic and prognostic purposes.

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