Case Report

Uterine neoplasm resembling an ovarian sex cord tumor

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ABSTRACT

Uterine neoplasm resembling an ovarian sex cord tumor is a rare neoplasm. Patients are generally of reproductive age, although a few may be perimenopausal or post menopausal. This neoplasm produces abnormal vaginal bleeding and uterine enlargement, suggesting leiomyoma or polyps. Based on the histology and clinical outcome, this tumor can be classified into 2 groups, namely one with focal areas resembling sex cord elements with a tendency to recur or metastasize and another with exclusive sex cord elements which runs a benign course. Immunohistochemical stains of the sex cord elements may show positively for vimentin, cytokeratin, actin and desmin in variable proportions. Inhibin is a more specific marker for these cells. Our case was a 50-year- old Omani lady with menorrhagia. Histology revealed features consistent with uterine neoplasm resembling an ovarian sex cord tumor.

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Uterine tumors with foci simulating ovarian sex cord tumors were first described by Clement and Scully[1] and were named as uterine tumors resembling ovarian sex cord tumors. In their detailed study of 14 cases, the authors divided these tumors into 2 groups based on the relative proportion of sex cord like elements and endometrial stromal cells. Group one tumors showed features of an endometrial stromal tumor with focal areas resembling sex cord elements. These showed a propensity to recur or metastasize. Group 2 tumors had a predominant or exclusive pattern of sex cord like elements. These tumors followed a benign course. The nature of these sex cord like formations has been widely disputed. Some authors concluded that these formations were epithelial, whereas others suggested a myoid phenotype.[2] Some believe that they are truly of sex cord nature. It is possible that these sex cord elements are derived from uterine stroma which, is capable of divergent differentiation. One such case is being reported here due to its rarity and striking histological appearance.

Case Report. A 50-year-old Omani lady was admitted with menorrhagia of 9 months duration. Ultrasound examination revealed a small endometrial polyp. Uterus was normal in size. Adnexae were free. Dilatation-curettage and polypectomy were carried out, and the tissue was sent for histopathological examination. Microscopy showed endometrial stromal tissue mixed with anastomosing cords, broad trabeculae and nests of "epithelial like" cells. These cells were round to oval with vesicular nuclei, inconspicuous small nucleoli and scanty cytoplasm **(Figure 1)**. The tumor was not mitotically active. Endometrial currettings, which were sent separately, along with the polypectomy specimen, showed secretory glands and features of chronic endometritis. Immunohistochemical stains on the tumor showed positivity for vimentin, actin and focal cytokeratin. The "epithelial like" cells were positive for estrogen and progesterone receptor markers. Epithelial membrane antigen and desmin tested negative. A diagnosis of uterine neoplasm resembling an ovarian sex cord tumor was made. The patient was lost to follow-up.

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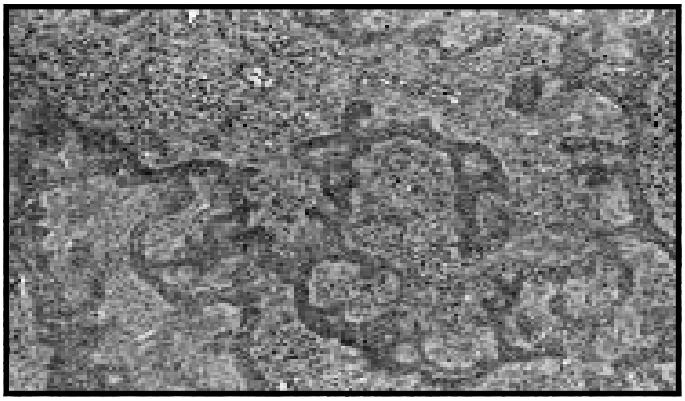

Figure 1 - Showing neoplastic cells arranged in cords and trabeculae. The stroma in between resembles proliferative endometrium.

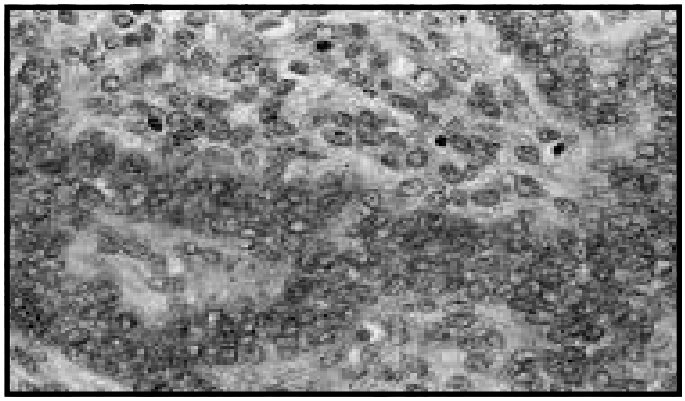

Figure 2 - High power view showing trabeculae composed of round cells with indistinct cell margins and round to oval nuclei.

One year later the patient was admitted again with menorrhagia. Ultrasound examination revealed a small polypoid lesion arising from the fundus. The uterus was normal in size. Fornices were free. Dilatation-curettage yielded fleshy fragments which on histology, appeared to be more cellular than the previous tumor, with mild pleomorphism. Mitotic activity was seen (3 per 10 high power fields). Small foci of necrosis were also noted.

Discussion. There are 2 uterine neoplasms that contain endometrial stroma that are variants of the usual endometrial stromal tumor. One is termed as uterine neoplasm resembling an ovarian sex cord tumor, and the other one, a combined smooth muscle stromal tumor. It was Clement and Scully[1] who first drew attention to the uterine neoplasm resembling an ovarian sex cord tumor. The neoplastic cells, which give the tumor its name, are arranged in cords, trabeculae, hollow tubules and sheets resembling epithelial cells, with scanty cytoplasm, indistinct cell margins and round nuclei.[2] The stroma in between these epithelial cells shows an appearance ranging from paucicellular to fibroblastic and sometimes simulating the compact stroma of proliferative endometrium. The growth pattern of the epithelial cells is reminiscent of ovarian sex cord tumors. Clement and Scully[1] in their analysis of 14 cases divided these tumors into 2 groups based on the relative proportion of sex cord like elements and endometrial stromal cells. Group one tumors contained focal areas resembling sex cord elements in an otherwise, typical endometrial stromal tumor. Group 2 tumors had a predominant or exclusive pattern of sex cord like elements. The biological behavior of these 2 tumors was different. Group one tumors showed a tendency to recur or metastasize, whereas Group 2 tumors followed a benign clinical course. Our case showed features consistent with Group one tumors. Numerous studies have been conducted to confirm the histogenesis of the sex cord like elements. No definite agreement has been reached though epithelial, myoid and truly sex cord nature has been postulated. Krishnamurthy et al[3] studied 7 examples of Type II tumors with markers of sex cord, steroid cell differentiation or both (inhibin, O_{13} and A_{103}.)[4] One or more of these markers were positive, in addition to constant immunoreactivity for vimentin and hormone receptors and variable positivity for keratin, actin and desmin. This provided strong evidence of true sex cord elements in this tumor, the importance of which remains speculative. Baker et al[5] studied staining of Inhibin, which is a peptide hormone expressed by normal ovarian granulosa cells and ovarian sex cord neoplasms. Inhibin staining was confined to the epithelial elements, which again confirmed the findings of Krishnamurthy et al.[3] In our case vimentin, actin, estrogen and progesterone receptors were strongly positive. There was focal cytokeratin positivity. Hence, it was diagnosed as uterine neoplasm resembling an ovarian sex cord tumor (Group one). This type has a tendency to recur or metastasize. One year later, our patient again presented with menorrhagia. Dilatation-curettage revealed a similar tumor, which appeared to be more cellular, but had similar trabeculae of epithelial cells resembling sex cord elements as before. Mitotic figures were present, upto 3 per 10 high power fields. Areas of necrosis were also seen. Since this is a rare tumor, not much data is available regarding the behavior of these neoplasms. However, the same criteria may be used to distinguish benign from malignant sex cord tumors as in stromal nodule versus low grade stromal sarcoma. Only those tumors with circumscribed borders, low mitotic index and uniform cells are considered to be benign.[4] In this case, the recurrent tumor was more cellular with mild pleomorphism and necrosis. Mitotic activity was also increased. Hence, it could be considered as a low-grade sarcoma. Malignant behavior is possible if the neoplasm is dominated by endometrial stroma and if it has an infiltrating margin.[6]

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