A puzzling ovarian tumour: pregnancy luteoma with diffuse endometriosis

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Key words
Ovary • Pregnancy • Luteoma • Endometriosis

Summary

Background.

Pregnancy luteoma is a distinctive non-neoplastic hormone dependent lesion arising in pregnancy and mimicking an ovarian tumour. Fewer than 200 cases have been described in the English-language literature. Its clinical and morphological features are characteristic and must be considered in order to prevent diagnostic misinterpretation.

To the best of our knowledge the association of pregnancy luteoma with endometriosis has not been reported in literature to date.

Case report.

A 30-year-old pregnant woman with no particular past medical history, consulted her gynaecologist at 17 weeks gestation for routine check-up. The patient was asymptomatic and did not show any signs of virilization. Ultrasonography disclosed a left adnexal heterogeneous mass measuring 7 cm in diameter with intramural vegetations. The right ovary was unremarkable. The patient underwent salpingo-oophorectomy considering the imaging findings were suspicious for malignancy. Histologically, the lesion was constituted of large sheets of luteinized polygonal cells with abundant eosinophilic cytoplasm and small round nuclei devoid of atypia and mitotic figures. In addition, there were several ectopic endometrial glands surrounded by abundant decidualized or edematous stroma. Immunohistochemically, these glands were immunoreactive for cytokeratin 7. The final pathological diagnosis was pregnancy luteoma associated with diffuse endometriosis.

Conclusions.

Because of its relative rarity, pregnancy luteoma is likely to be clinically misinterpreted and overtreated, as in the present case.

Introduction

Luteoma of pregnancy is a rare non-neoplastic hormone dependent lesion of the ovary that emerges during pregnancy and regresses spontaneously after delivery. It is usually asymptomatic and is found incidentally during a caesarean section or postpartum tubal ligation. Fewer than 200 cases have been described in the English-language literature to date. Clinical and morphological features of pregnancy luteoma are characteristic and must be considered in order to prevent diagnostic misinterpretation. In this paper, the authors report a particular and misleading case of pregnancy luteoma clinically misinterpreted as an ovarian neoplasm. Emphasis is placed on some unusual histological features namely the presence of endometriosis. To the best of our knowledge, the association of pregnancy luteoma with ectopic endometrial glands has not been reported in literature to date.

Case report

A 30-year-old pregnant woman (Gravida 1, Para 0) with no particular past medical history, consulted her gynaecologist at 17 weeks’ gestation for routine check-up. The patient was asymptomatic and did not show any signs of virilization. Physical examination did not disclose any anomaly. Ultrasonography showed a single uterine pregnancy with fetal cardiac pulsations. There was also a left adnexal heterogeneous partially cystic mass measuring 7 cm in diameter with intramural vegetations. On MRI, the heterogeneous nature of the tumour was clearly identified, with solid and cystic components. The tumour was characterized by low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. The right ovary was unremarkable. The patient underwent salpingo-oophorectomy considering the imaging findings were suspicious for malignancy.

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Macroscopically, the left ovarian mass had a smooth surface, measured 7 cm in its greatest diameter and was well demarcated. On the cut surface, it appeared partially cystic with several solid fleshy brown areas (Fig. 1a). Histologically, the solid areas were constituted of large sheets of luteinized polygonal cells with abundant finely granular eosinophilic cytoplasm and small round nuclei devoid of atypia and mitotic figures (Figg. 1b, 1c-1d). In addition, there were several ectopic endometrial glands surrounded by abundant decidualized (Figg. 2a-2b) or edematous stroma (Figg. 2c-2d). Immunohistochemically, these glands were immunoreactive for cytokeratin 7 (Fig. 3). No necrotic areas or typical Reinke’s crystalloids were identified. The final pathological diagnosis was pregnancy luteoma associated with diffuse endometriosis. Twenty-three weeks later, the patient spontaneously delivered a girl with no signs of virilization. At present, the patient is well and is still being followed up.

Discussion

Between 1% and 2% of pregnant women have an adnexal mass that is sonographically detected. Common adnexal lesions associated with pregnancy include simple cysts, hemorrhagic cysts, leiomyomas, and hyperstimulated ovaries in patients who have undergone assisted fertility. Uncommon adnexal lesions specific to pregnancy include hyperreactio luteinalis, and luteomas. Adnexal masses associated with pain include ovarian torsion and heterotopic pregnancy. Some adnexal entities are found incidentally, such as teratomas, endometriomas, hydrosalpinx, cystadenomas, and cystadenocarcinomas. Pregnancy luteoma is a rare non-neoplastic lesion characterized by ovarian enlargement during pregnancy. It usually occurs in the third and fourth decades of life and about 80% are associated with multiparous women. Our patient was 30 years old gravida 1, para 0. Pregnancy luteomas are estimated to be bilateral in one third of the cases and multinodular in half of the cases. In our case, the lesion was unilateral. Pregnancy luteomas are...
thought to arise because of excessive response of stromal cells to pregnancy hormones, especially hCG. However, luteomas are not commonly seen in conditions that result in marked elevation of hCG, such as trophoblastic disease, hyperreactio luteinalis, or early in pregnancy, suggesting other unknown underlying or contributing factors in their development. Luteomas cause maternal virilization in 25% to 30% of cases and carry a 50% risk of virilizing a female fetus. In our case, neither maternal nor fetal virilization were noted. Pregnancy luteomas are often asymptomatic and are usually discovered incidentally at the time of the caesarian section. Our patient was asymptomatic. Given the size of some of these tumours, they may be associated with mass effect on adjacent organs like the ureter, presenting with symptoms of obstructive uropathy or are complicated by infarction, which can lead to ovarian scarring. They can rarely be associated with torsion and present with severe abdominal pain. The tumours can rupture, leading to massive blood loss, hence necessitating surgical management. There have also been rare reports of pregnancy luteomas presenting like malignant tumours with massive ascites and raised CA-125. Investigations must be
minimally invasive with no significant detrimental effects to the developing fetus and imaging modalities like MRI and ultrasonography have shown promise in being able to help diagnose the tumor and prevent unnecessary interruption of pregnancies. On ultrasonography, luteomas appear as heterogeneous solid masses, predominantly hypoechoic compared with normal ovarian tissue, with thick walls and irregular internal contours in an enlarged ovary. They are often highly vascular and mimic ovarian neoplasms. MRI gives different results in virilizing and non-virilizing tumor, as virilizing tumors often show relative high signal intensity on T1-weighted images and low signal intensity on T2-weighted images (due to the colloid material in the cells), while other neoplastic solid tumors show characteristic low signal intensity on the T2-weighted images. When a luteoma is suspected, laparotomy can be avoided during pregnancy because the lesions resolve by 2 to 3 weeks post partum. Surgery is only required for those with an acute presentation and when differentiation from malignancy is not possible based on imaging. Pregnancy luteomas are variable in size, ranging from microscopic to over 20 cm in diameter. On gross examination, cut surfaces of luteomas are solid, soft, tan or flesh colored, with hemorrhagic foci. Microscopic examination discloses the presence of sharply circumscribed rounded masses of cells with follicles containing pale fluid or colloid material. These cells are likely to originate from ovarian stromal cells or internal theca cells as a response to high levels of hCG. They are round to polygonal with abundant, finely granular, eosinophilic cytoplasm and small nuclei. The presence of both luteoma and a granulosa cell proliferation in a Sertoli-like pattern as well as ectopic decidua have been reported by some authors, however ectopic endometrial glands could be confused with primitive or metastatic adenocarcinoma or entrapped follicles, however there were no cytological atypia and the glands were immunohistochemically positive for CK7.

In conclusion, pregnancy luteoma is a tumour-like lesion which can mimic an ovarian neoplasia. Although the diagnosis of pregnancy luteoma relies on the clinical setting and histological grounds, it must be presumed in pregnant women with an ovarian unilateral or bilateral mass. Because of its relative rarity, it is likely to be clinically misinterpreted as in the present case. The physiological regression of pregnancy luteoma soon after delivery allows a conservative approach.

References