Carcinosarcoma arising in a dermoid cyst of the ovary

D S Arora, S Haldane

Abstract

A case of carcinosarcoma arising within an otherwise benign cystic teratoma is reported. The patient, a 78 year old nulliparous woman, presented with right sided abdominal pain of short duration and subsequently underwent a bilateral salpingo-oophorectomy. Slicing of the left ovary revealed a unilocular cyst containing hair admixed with soft yellow material with a thin wall apart from a solid area at one pole. Extensive areas of necrosis and cystic degeneration were present within this mass. Histologically, the large cyst was a typical mature cystic teratoma, containing carcinomatous and sarcomatous elements. Mature cystic teratomas have been reported in association with a variety of malignant ovarian tumours such as mucinous cystadenocarcinoma and malignant germ cell neoplasms. Secondary malignant transformation within a dermoid cyst is a much rarer occurrence, estimated as less than 2% of all such lesions. Adenocarcinomas are the second most common malignancies arising within dermoid cysts. Sarcomas alone or in combination with squamous carcinoma have been described arising in a mature cystic teratoma. To the best of our knowledge, no case of sarcoma arising in association with adenocarcinoma has been described before.


Keywords: ovary, dermoid cyst, carcinosarcoma.

Malignant transformation within a mature cystic teratoma (dermoid cyst) occurs in less than...
2% of cases' and most commonly results in a squamous cell carcinoma (75–85%). Other reported malignancies include adenocarcinoma (7%), various pure sarcomas (7%), melanomas and basal cell carcinomas (each less than 1%), and anaplastic carcinoma. Here, we describe a case of carcinosarcoma arising within an otherwise benign cystic teratoma which, to our knowledge, is the first reported case of such a combination.

Case report
A 78 year old nulliparous women presented with right sided abdominal pain of short duration. On examination, a large mobile mass was present in the lower abdomen. Ultrasound showed a partly solid and partly cystic ovarian mass. The patient underwent a bilateral salpingo-oophorectomy two days after admission.

Pathological findings
The left ovary was enlarged and measured 23 × 17 × 15 cm. Slicing revealed a unilocular cyst, 14 cm in diameter, containing hair admixed with soft yellow material with a thin wall apart from a solid area at one pole, measuring 10 × 8 × 7 cm. Extensive areas of necrosis and cystic degeneration were present within this mass. The right ovary and both fallopian tubes appeared normal.

Histologically, the large cyst was a typical mature cystic teratoma lined by squamous epithelium and adnexal structures (fig 1 inset). The solid component consisted of glandular structures set in an abundant cellular stroma. Glands showed both secreted and intracellular periodic acid Schiff positive diastase resistant mucin, and varied in architecture from rounded acinar structures to poorly differentiated signet ring cells (fig 1A). The stroma consisted of uniform plump mesenchymal cells with high nuclear cytoplasmic ratios (fig 1B). Some smooth muscle differentiation was present, but no differentiation towards any other mesenchymal elements was identified. The carcinomatous and sarcomatous elements were closely associated and intermixed with each other. Mitoses were abundant in both the glandular and stromal components, including some abnormal forms. There were extensive areas of necrosis. No benign glandular epithelium was present.

Immunohistochemical findings
The glandular component stained positively for carcinoembryonic antigen (Dako, Glostrup, Denmark) and CAM 5.2 (Becton Dickinson, California, USA). The malignant stromal component was positive for vimentin and showed mild positivity for α smooth muscle actin (Sigma, Poole, Dorset, UK), and equivocal staining for desmin (Dako). Neither component stained positively for S-100 (Dako), neuron specific enolase (Dako), chromogranin (Dako), or synatophysin (Dako).

Discussion
Mature cystic teratomas have been reported in association with a variety of malignant ovarian tumours such as mucinous cystadenocarcinoma and malignant germ cell neoplasms. Secondary malignant transformation within a dermoid cyst is a much rarer occurrence, estimated as less than 2% of all such lesions. Table 1 shows the variety of reported tumour types in this situation.

We are confident that our case presented with primary ovarian disease, as there was no evidence of any other primary site and the tumour was confined to the left ovary. The striking production of neutral mucins, glandular architecture and foci of signet ring morphology suggest expression of an upper gastrointestinal phenotype, although no corresponding benign glandular epithelium was identified. The stroma consisted of uniform plump cells with a high nuclear:cytoplasmic ratio and with focal expression of smooth muscle antigens but no differentiation towards other mesenchymal phenotypes.
Malignant dermoid cyst of the ovary

Our case must be distinguished from malignant mixed müllerian tumour which occurs in the same age group and has carcinomatous and sarcomatous components. However, these tumours are generally solid, lack the characteristic squamous epithelial lined cyst and show different mucin expression. Immature malignant teratoma occurs in a younger age group and shows a variety of lines of mesenchymal differentiation, notably towards neural elements, which were absent in this case.

Germ cell tumours of all types derive from neoplastic totipotent stem cells whose progeny retain the ability to differentiate towards tissues of all three embryonic somatic layers and the transitional extraembryonal structures. The original pattern of the better differentiated members of this group suggests that local cellular influences are important in determining the final phenotype expressed by individual cells. The histological behaviour and histological appearances of these tumours are determined by the degree to which this embryogenetic environmental responsiveness is retained. In contrast, our case resembles carcinomas at other sites in that there was an essentially random mixture of stromal and epithelial elements. This reflects a probabilistic commitment of individual cells and clones to one or other expressed phenotype without regard to organogenetic influences. Our case implies glandular malignant transformation within the previously benign, fully differentiated, glandular component of a mature cystic teratoma, although no premalignant changes were identified despite thorough sampling.

Adenocarcinomas are the second most common malignancies arising within dermoid cysts and may display gastrointestinal phenotypes, but lack the malignant stroma as in our case. Sarcomas alone or in combination with squamous carcinoma have been described arising in a mature cystic teratoma. To the best of our knowledge, no case of sarcoma arising in association with adenocarcinoma has been described before.

The prognosis of patients with malignant transformation in teratoma is very poor, with most women dying within one year. Occasional cases with prolonged survival have been reported. Poor prognostic factors include tumour dissemination, cyst wall invasion, ascites, spontaneous or accidental rupture, adhesions, and tumour type other than squamous carcinoma. Treatment is surgical, with pelvic clearance in most cases. Chemotherapy may be indicated according to the tumour type.


Tenascin in human papillomavirus associated lesions of the uterine cervix

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Abstract
The immunohistochemical expression of tenascin was studied in 80 morphologically diagnosed condylomas and cervical intraepithelial neoplasia (CIN) lesions. The results were compared with the human papillomavirus (HPV) DNA subtype, which was determined by HPV dot blot and in situ hybridisation. Tenascin mRNA synthesis was also determined in 10 selected cases by in situ hybridisation. No statistically significant association was found between tenascin expression and the degree of dysplasia or the HPV subtype. There was, however, a strong correlation between the extent of tenascin immunoreactivity and the degree of inflammation. Synthesis of tenascin mRNA was detected in basal keratinocytes and in fibroblasts by in situ hybridisation. The lack of association between the grade of CIN and tenascin expression precludes its use as a marker of premalignancy in CIN.

Keywords: tenascin, extracellular matrix proteins, human papillomavirus infection, HPV DNA, inflammation.

Human papillomavirus (HPV) is the aetiological agent of various genital lesions and there is
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