# SARCOMA BOTRYOIDES OF THE CERVIX

## A CASE REPORT AND REVIEW

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Sarcoma botryoides is usually regarded as one variety of the rare group of mesodermal mixed tumours of the female genital tract. It occurs most commonly in infants and children under the age of 2 years, and in the vast majority arises primarily from the vaginal subepithelial tissues. In women in the reproductive age sarcoma botryoides is much rarer and tends to originate in the cervix, in the form of soft polypi, which are sometimes macroscopically benign and even on histological examination may not reveal any evidence of malignancy. The malignant nature of these polypi then becomes evident only when they recur after removal. A case of sarcoma botryoides of the cervix which presented in this manner, is described.

### CASE REPORT

Mrs. A. C. de K., a White woman aged 45 years, para 4+1, was first seen in February 1963, complaining of slight intermenstrual bleeding for 6 months and of something protruding at the vaginal introitus for 1 month. She had a regular 5/28-day cycle. Apart from stress incontinence, there were no other gynaecological complaints. Her previous history was negative. Apart from the fact that her mother had diabetes mellitus, the family history was also negative. On general and systemic examination no abnormalities were found. On genital examination, the vulva, perineum and urethral meatus were normal. There was a greenish-yellow vaginal discharge and a vaginitis clinically suggestive of infection with *Trichomonas vaginalis.* One large and several smaller pinkish, pedunculated polypi were seen to arise from the endocervix. They were soft in consistency and had no features suggesting malignancy. The uterus was slightly bulky in size, situated in the axis of the pelvis, mobile and non-tender. No abnormality could be palpated in the adnexa. There was a grade 1 uterine prolapse with a slight cystocoele and moderate rectocoele, and no demonstrable stress incontinence. Papanicolaou smears revealed class II cytology, with evidence of cervicitis and trichomonas vaginitis and hyperplastic adenomatous cells. The haemoglobin estimation was 14 G/100 ml. and the urine contained no abnormal constituents.

The patient was admitted, and a dilatation and curettage, polypectomy and cauterization of the cervix were carried out under general anaesthesia. Histological examination revealed cervical polypi covered partly by squamous and partly by columnar epithelium, with slight epidermidization of the cervical glands, and secretory endometrium. The stroma was very oedematous and contained numerous chronic inflammatory infiltrations. No anaplasia or other signs of malignancy were observed.

In July 1963, 5 months later, the patient was readmitted with the complaint that polypi were again visible at the introitus. She also complained of a vaginal discharge and stated that on one occasion she had passed some fleshy material *per vaginam* followed by bright red bleeding. Examination now revealed a cluster of grapelike endocervical polypi, which bled to the touch. The rest of the findings were the same as at the first admission. The cytological report on cervical and vaginal smears was now class III minus, with groups of adenomatous and pleomorphic cells, but no definite evidence of malignancy.

The patient was admitted forthwith, and a polypectomy was again performed. Histological examination showed polypi with squamous and columnar epithelium and chronic inflammatory infiltration. The stroma was oedematous in parts and in contrast to the first specimen the connective tissue was cellular in some areas. As the stromal cells, however, were normal in shape and size, the cellularity was not considered as a definite sign of malignancy.

In view of the age of the patient and the rapid recurrence of the polypi, however, a vaginal hysterectomy, anterior colporrhaphy and posterior colpoperineorrhaphy were performed. Histological examination of the uterus revealed chronic cervicitis and signs of a recent excision as the only abnormality.

The patient made an uneventful recovery and was discharged home on the 8th postoperative day. At the follow-up examination 6 weeks later the patient had no complaints. A few granulations at the vault of the vagina were cauterized. She was seen again a month later; the granulations at the vault had healed completely and she was finally discharged.

The patient remained perfectly well until July 1965, when she presented again with postcoital bleeding and a feeling of discomfort in the vagina for 1 month. General and systemic examination were quite negative. On vaginal examination, a bloodstained discharge was present. In the vault of the vagina 3 rounded, smooth, soft polypi, each about 3 cm. × 1 cm., were found. One was dark red and haemorrhagic, but the other two were pale and nearly transparent. On bimanual examination a soft, rounded, smooth, well-circumscribed mass about 7 cm. (3 inches) in diameter, was palpable above, and attached to, the vaginal vault. It extended towards the left lateral pelvic wall, but was not fixed to it. The bladder and rectum did not appear to be involved. The mass was so soft as to suggest an analogy to liquid encased in a cellophane bag.

Cytological examination revealed a class V, with frankly malignant cells, which were however so undifferentiated that their origin could not be determined.

The patient was again readmitted. Cystoscopy and radiological examination for metastases were negative. The polypi were removed and histological examination now left no doubt as to the nature of the tumour: Histological sections revealed polypoid tumours which contained no glands and were covered by squamous epithelium. The stroma was oedematous and infiltrated by inflammatory mononuclear cells. The tissue was cellular and showed dense perivascular accumulations of connective tissue cells as are frequently seen in sarcomata. The nuclei were larger than normal and hyperchromatic. Cellular pleomorphism was not marked. In places the tissue looked myxomatous. A diagnosis of a mixed mesodermal tumour was considered and sections were stained with Mallory's PTAH which showed a few striated muscle fibres. The slides made from the polypi removed on the previous two occasions were reviewed, and in the polypi removed before hysterectomy, rhabdomyoblasts were now also found (Fig. 1). The patient

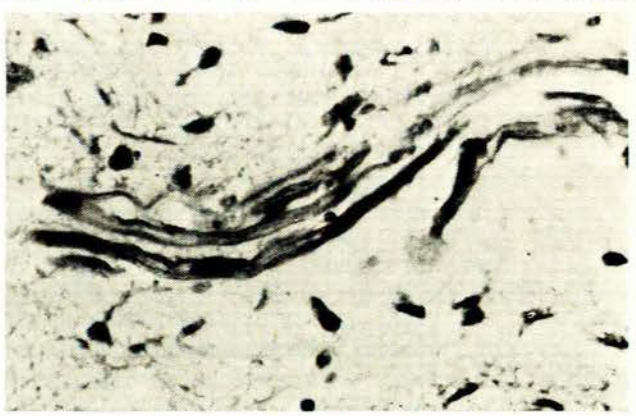

*Fig. 1.* See text.

was given telecobalt therapy, at a tumour dose of 200r/day, to a total of 6,000r. The tumour at the vault of the vagina disappeared completely, and when last seen, 9 months later, she was still free from recurrence.

## DISCUSSION

Mesodermal mixed tumours of the uterus and vagina, although not as uncommon as was previously thought, are still rare. Marcella and Cromer,[1] reporting 11 cases, could find only 234 cases in the literature up to that time (1959). Since then, in two large series in the American literature,[2,3] 29 cases of mesodermal mixed tumours are reported among 87 cases of uterine sarcoma. Sarcoma botryoides, recognized as a distinct variant of mixed mesodermal tumours because of special clinical and pathological features, is, again, an uncommon variety of these tumours. Thus, in the two large series referred to above, there are only 4 cases of sarcoma botryoides (although White *et al.*[3] state that 6 cases of sarcoma botryoides occurring in children are excluded from their series of uterine sarcoma) and Taylor[4] described 7 cases among 40 mesodermal mixed tumours. The vast majority of cases of sarcoma botryoides occur in infants and children, and arise primarily in the vagina. Thus, among the 13 cases described by Daniel *et al.*[5] they all appeared to have their origin in the vagina, although the cervix was secondarily involved in some cases. Sarcoma botryoides may be multifocal in origin, however. During the reproductive years, the cervix is the predominant site of origin. Simpson[6] in 1943 could collect only 22 of these tumours of the cervix from the world literature, with an average age of 34 years. Most of the cases reported since then have been vaginal, although Taylor[4] described 4 cervical cases, in patients aged 7 months and 20, 38 and 48 years respectively. In some cases[2,3] the site of origin is not clearly stated.

### *Clinical Features*

Sarcoma botryoides presents most often with abnormal uterine bleeding, or with a mass protruding from the introitus, or with the history of having passed some tissue *per vaginam*. All three of these features were present in our case. The tumour consists of multiple, soft, pinkish semitranslucent gelatinous polypi, first likened to a bunch of grapes by Pfannenstiel (1892) ('Traubiges sarcom').[7] It has also been likened to the vesicles of a hydatidiform mole. The polypi often appear clinically benign, and even the pathologist may be initially deceived, as in our case. Thus Amolsch,[8] for instance, in reporting 6 cases of mesodermal mixed tumours of the uterus and vagina, states that the initial diagnosis in all was a benign polyp. These polypi then show a marked tendency to recurrence after local removal. It has now become widely recognized that benign polypi in children are extremely rare, and that sarcoma botryoides is the commonest malignant tumour of the genitalia in children; hence any vaginal polyp in a child must be regarded with the greatest suspicion.[9]

### *Histogenesis and Histopathology*

The specimens of this patient gave us the opportunity to study the development of a malignant mixed mesodermal tumour from an innocent-looking polyp to a full-fledged sarcoma. The polypoid tissue (in all specimens) was very oedematous. Whereas the connective tissue of the first biopsy was of normal cellularity, there was an increased cellularity in the second biopsy. This was interpreted as actively growing connective tissue which is occasionally found in cervical polypi. Striated muscle fibres were originally not observed and therefore the lesion was considered as probably benign without definite signs of malignancy.

About two years after this biopsy was taken, the last specimen from this patient left no doubt as to its malignancy. Stellate cells were abundant and myxomatous change and striated muscle fibres were found.

In retrospect, the histological findings demonstrate the gradual development of histological signs of malignancy and give some indications concerning the pathogenesis of these tumours. These indications are the progressive increase in cellularity, the later appearance of anaplasia, and the presence of striated muscle fibres in the second and third specimen but not in the first. Therefore, Wilms' theory that these neoplasms are teratomatous can be discounted. It seems much more probable that these tumours arise either from pluripotential mesenchymal cells which differentiate in different directions or from stromal cells which undergo metaplasia as originally suggested by Pfannenstiel.[7]

### *Treatment and Prognosis*

Marcus,[10] in reporting 2 cases of sarcoma botryoides of the cervix—one of which had survived for 7 years at the time of reporting, could find only 5 previously reported 5-year cures in the literature. The modern tendency is to favour radical surgery in sarcoma botryoides of the vagina[5,11] in the belief that the tumour tends to spread by local infiltration in the first instance. Sarcoma botryoides of the cervix also gives rise to lymph node metastases quite frequently, and pelvic and even inguinal lymphadenectomy is thus also advised.[1,10] Two of the 6 survivors for 5 years or more referred to above, however, were treated merely by local excision and radiotherapy. No 5-year cures have been achieved by means of radiotherapy alone in cases of sarcoma botryoides, but Taylor[4] reports a patient with carcinosarcoma of a cervical stump surviving for 6 years with radium treatment only. The dramatic response in our patient to telecobalt therapy would tend to lend support to Taylor's opinion that 'it would seem unwise to accept the statement that radiotherapy has no place in treatment'.

The use of chemotherapy in the form of alkylating agents, antimetabolites and the antibiotic, actinomycin D, has been reported by a number of authors,[1,3,5,12,13] with, at best, only very temporary remission. Hreshchyshyn and Holland[13] state that recent personal experience suggests that actinomycin D is active against mixed mesodermal tumours.

## SUMMARY

A case of sarcoma botryoides of the cervix presenting as recurrent cervical polypi, is presented. Dramatic response to telecobalt therapy has been obtained. The incidence, clinical features, histogenesis, histopathology, treatment and prognosis are briefly discussed.

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