SARCOMA BOTRYOIDES

(A Case Report)

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Botryoid sarcoma occurs in infancy and childhood and has also been reported in a newborn child. It was first described by Spiegelberg (1879) as sarcoma colli uteri hydropicum papillare. The term, sarcoma botryoides first suggested by Pfannensteil (1892), was derived from the Greek term 'Botpus' meaning a bunch of grapes and refers to the naked eye appearance of the tumour. Kehrer (1906), and Meikle (1936) included it under the title of mixed mesodermal tumours because it often contains a variety of tissue heterotopic to the location in which it is found. However, this tumour is monodermal in origin and should not be confused with teratomas. Botryoid tumours contain a variety of tissues including striated muscle fibres and their embryonic forms, myxomatous tissue, cartilage, osteoid tissue, smooth muscle fibres, adipose tissue, lymphomatous elements and a mixture of cellular, myxomatous and embryonic forms of sarcoma. A case of such a grape-like growth arising from the vagina and cervix in a girl, two and a half years old, is reported below.

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Case Report

Patient two and a half years old, was admitted on 10th June, 1966 with complaints of (i) a growth coming out of the vagina for twelve days, (ii) burning sensation during micturition for twelve days and (iii) fever off and on for 12 days. The mother noted the rise of temperature and the growth in the vagina and that the child used to cry while passing urine. She belonged to a very low-socio-economic stratum. All her brothers and sisters were well. On examination the child was found to have an average build with no pallor, no cyanosis and no oedema. Cardiovascular and respiratory systems were normal. Palpation of abdomen did not reveal enlarged liver or spleen and the lower abdomen was soft and without tenderness. Local examination of the external genitalia revealed a growth, resembling a bunch of grapes, coming out of the vagina.

Total white blood cell count was 12,000/cmm and differential count showed a relative lymphocytosis (lymphocytes 48%, polymorphs, 44%, eosinophils 5% and monocytes 3%). The haemoglobin value was 11 gms%. Stool examination showed no ova or cysts. Routine urinalysis showed acid reaction, deep yellow colour, albumin and sugar in traces, large number of oxalate crystals and occasional pus cells.

On examination under anaesthesia the growth was seen coming out of the vagina which had a typical bunch of grapes appearance (Fig. 1); the pedicle of the growth

could not be reached and therefore, only a portion of the growth was excised and sent for histopathology. Vaginal examination with the little finger revealed that the whole of the vagina was filled with the tumour. Rectal examination showed that the rectum was quite free from the surrounding tissue.

Histopathology report

Section showed transversely cut small polypoidal masses of connective tissue with overlying strips of stratified squamous epithelium. The connective tissue cells were characteristically long and spindle-shaped with fine tapering processes and with abundant oedema and inflammatory changes comprising newly formed capillaries and infiltration by polymorphs and few eosinophils. The connective tissue cells showed mitotic figures indicating active proliferation, but no clear-cut evidence of malignancy. The pathologist commented that the tissue represented actively proliferating connective tissue overgrowth without histological evidence of malignancy, but advised that the patients should be carefully followed up.

On the basis of this histological report the patient was given a course of penicillin and streptomycin (Dicrysticin Pediatric) for 10 days and discharged with instructions to attend the out-patient's department every fortnight for check up. But this instruction was not followed.

The girl was again brought by her parents on 19th July, 1967 (about a year after her discharge from hospital) with the complaints of a growth in vagina and blood-stained, foul smelling dirty discharge for one year. On examination the girl showed marked pallor and weakness. Regional lymph nodes were not enlarged. Her haemoglobin value was 9.6 gms.%. Since the patient had fever and purulent vaginal discharge she was given a course of penicillin streptomycin injections for a week.

When the fever subsided and the vaginal discharge cleared up a Schuchardt's incision was made under general anaesthesia and the vagina was explored. It was found to be balooned out. There was a cystic mass arising from the left fornix. The cervix was pushed on the right side and covered with a polypoidal growth. The uterine body could not be felt. An elongated mass was felt high up in the right iliac fossa. The base of the growth was clamped and cut. After the removal of growth, the uterus was felt to be pushed towards the right side. The Schuchardt's incision was closed in layers. The patient stood the operation well. The growth was sent for histopathology.

Histopathological Report

Section showed fibromyomatous polypoidal growth with abundant oedema in which thin long strips of embryonal striated muscle fibres were noticeable. There was a regular thin layer of stratified epithelium covering. There were no features to show distinct malignancy. Most nuclei and some of the connective tissue tumour cells presented some enlargement and some hyperchromaticity. The picture was absolutely identical to what was observed a year ago from the same patient. In the absence of truly sarcomatous pattern of the grape-like sarcoma arising from the vagina in children the picture could be best interpreted as its benign counterpart.

The patient was discharged after 15 days of the operation and was advised again to attend out-patient deparment every fortnight for follow-up. But the patient was brought again after 6 months. The growth had recurred and the patient had grown anaemic and cachectic. Her condition gradually deteriorated and she ultimately died of cachexia and intercurrent infection about a year after the operation.

Discussion

It is a very rare and highly malignant tumour and occurs predominantly in infants and children, although it has been known to occur in patients of all age groups. Most common site of origin is the urogenital system, but it may involve other organs also. In infants the tumour arises from the vagina and vulva. In women of reproductive age, the tumour may be found to arise from the vagina, cervix or the body of the uterus. In post-menopausal women, such a tumour arises most often from the body of the

uterus. According to Willis (1953) mixed mesodermal tumours of the uterus fall into two distinct subgroups as regards age and site of origin, though generally similar in structure and behaviour. Most of the cervical growths, usually arising by a pedicle from the tip of the cervix, occur in young and adult women with a fairly uniform distribution over the first four decades. The corporeal growths having a broad base and arising from the fundus of the uterus are most commonly encountered in post-menopausal women.

In children the growth usually arises from the vulva, vagina or the portio vaginalis of cervix. The cervix may frequently be involved secondarily. The characteristic grape-like mass of pinkish oedematus polyps protruding from the vagina is almost pathognomonic (Daniel, *et al,* 1959). In advanced cases the whole of the vagina may be filled with the growth with extension to the cervix, uterus, parametrium and abdomen. Often, a precise origin is difficult to demonstrate, specially in the tumours of infancy, as they tend to fill the whole vagina causing it to baloon out. In some cases it may arise from the cervix also (Shaw, 1928; and Duncan and Fahmy, 1963). Daniel (1959) points to a multicentric origin giving rise to multiple polyps springing up at various levels of the vagina.

Histological examination reveals considerable variation in structure in different parts of the tumour as well as different tumours. The botryoid growth usually consists of loose myxomatous tissue covered by epithelium which is stratified squamous in vaginal and cervical growths and columnar in those arising from the uterine body. Groups of embryonic mesodermal cells are scattered in the stroma. Islands of cartilage of immature or adult type are often encountered. In corpus tumours the main component is sarcoma (interspersed with myxomatous tissue), with cell pleomorphism and nuclear irregularities. The glandular epithelium may be benign or malignant. Very rarely, gross striated muscle fibres may be seen. Epithelial tissue may be found in the substance of the growth and is frequently hyperplastic or carcinomatous, an example of a composite nature of the growth leading to the name carcino-sarcoma.

The spread of the tumour occurs by direct extension and distant metastases take place through lymphatics and blood stream. The treatment of the growths arising from cervix is Werthiem's hysterectomy and radiotherapy and for growths from the body of the uterus, panhysterectomy. Radical surgical extirpation is also necessary in case of growths arising from vagina and vulva often necessitating diversion of the urinary stream. However, even with radical surgery the chances of recurrence are very high (Dunster and Benett, 1953; and Taylor, 1958). Radiotherapy alone is almost always unsuccessful. Cases of long term survival after surgery have been reported by Ufelder & Quan (1947) and Cywes and Louw (1961); most of these cases had had exenteration with the implantation of the ureters into the intact colon or into a colostomy or into a segment of a bowel made as an artificial bladder. These operations have also been performed in children and those who have done so are convinced that they are worth doing, as with love and affection, these children are perfectly capable of overcoming the physical deformities inflicted upon them (Daniel *et al,* 1959; and Cywes and Louw, 1961).

As regards histogenesis there are many views. Wilms (1900) suggested that this

growths arose from misplaced embryonic rests carried down by the Wolffian duct or Mullerian system. Willis dismissed the embryonic cell rest theory as absurd on the ground that there were no chances of such undifferentiated cell rests surviving in the endometrium of a woman through multiple pregnancies and eventually producing a mixed heterotopic tumour at the age of 50, 60 or 70 years.

Pfnnensteil suggested that these growths arose from the endometrial stroma and several heterotopic tissues developed by aberrant differentiation or metaplasia. The well known plasticity o the cells of the endometrial stroma and their comparative primitiveness tend to prove the stromal metaplasia theory.

Sternberg (1954) suggested that these tumours arose from the Mullerian mesoderm and were monodermal in origin. Taylor (1958) emphasised that elements of stromal (Mullerian) origin with abnormal differentiation were responsible for bone and cartilage formation in the tumour. This theory of histogenesis of mixed mesodermal tumours is widely accepted.

Summary

A case of grape-like tumour from cervix and vagina is reported in a girl aged two and a half years. The histopathological reports, on both biopsy and the excision of the tumour, were not suggestive of malignancy. Therefore, no radical surgery was done. The patient died within 2 years of the appearance of the symptoms, indicating thereby the malignant nature of the growth.

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See Fig. on Art Paper I