Sarcoma Botryoides

—— Report of Two Cases ——

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Sarcoma botryoides, one of rare mesenchymal tumors, grows in the form of polypoid grapelike masses and derives its name from this gross appearance. Although most of the cases are located in the urogenital tract, neoplasms with similar gross and microscopic characteristics have been also reported to occur in the nasopharynx[1], the palate[2], the common bile duct[3)4)] and the anus[5].

The purpose of this paper is to report two cases of sarcoma botryoides, of which one arised in the cervix and the other appeared in the soft palate. These two cases are noteworthy in that the patients are still alive and free from recurrence of the tumor.

REPORT OF CASES

Case 1.

A 5-year-old girl was well until ten days before the admission when her mother first noted a polypoid mass of small egg size, protruding from the soft palate. The patient had suffered from difficulty in swallowing and speaking for two weeks.

At the end of December 1965, she admitted to Yamaguchi University Hospital with a chief complaint of painless tumor mass in the oral cavity. On admission, the patient was a well-developed and well-nourished girl. Physical examination disclosed no abnormalities except for a mass with soft consistency protruding from the soft palate. The tumor was grayish white and was small egg in size, almost filling the oral cavity. The surface was smooth and rather gelatinous.

Examination of the peripheral blood revealed 416×10^4 red blood cell and 7700 white blood cell, with a differential count of 62 per cent segmented

neutrophils, 23 per cent lymphocytes, 11 per cent monocytes, 3 per cent eosinophils and 1 per cent basophils. Blood chemistry was consisted of the follwing: Hb. 12.6 g dl, Serum protein 7.2 g/dl, A/G ratio 1.06, Albumin 3.7 g/dl, Globulin 3.5 g dl, Blood sugar 96 mg dl, Icteric index 4, Alk. phosphatase 3.7 u, Cholinesterase 0.47 △pH, Cholesterol 118 mg/dl, Phenol turb. test 13 u, GPT 2 u, LAP 15 u, LDH 36 u. Serological examination for rheumatic disease was negative. Wasserman reaction was negative.

A biopsy of the tumor resulted as sarcoma botryoides.

The lesion was treated by irradiation with use of Betatron and anticancerous drug (Endoxan) was also administered by intravenous injection. They were markedly effective and remarkable diminution in size of the mass was noted. In May, 1966, wide excision of the tumor tissue was performed under general anesthesia.

Post operative course was uneventful and she discharged at the beginning of July, 1966. She is now leading a normal life for a child of her age with no evidence of recurrence and metastasis of the tumor.

Histopathological Findings :

The tumor mass was covered by the normal stratified squamous epithelium (Fig. 1), but occasional areas of erosion were observed. Tumor was moderately vascular and acellular myxomatous area was prominent (Fig. 2). Around the blood vessel, tumor cells were more compactly arranged (Fig. 3). In general, tumor cells were spindle in shape and had hyperchromatic nuclei with prominent nucleoli (Fig. 4). Nuclear pleomorphism was prominent and mitotic figures were frequently visible. Though the cytoplasm of these cells formed delicate cytoplasmic projections, cross striations were not demonstrable in any of these cells by van Gieson stain or phosphotangustic acid hematoxylin (Fig. 5). Reticulum fibers were abundantly seen among the individual tumor cells and no alveolar structure was observed (Fig. 6).

Case 2.

The patient was 16-year-old high school girl who first noted stinking serosanguineous vaginal discharge at the end of May, 1964. About one month later, polypoid mass protruding from the vagina was noticed. Pulling down of the tumor mass by herself caused marked vaginal bleeding and she was promptly referred to a hospital. A biopsy of the tumor revealed sarcoma botryoides and she was admitted to the gynecological clinic of Yamaguchi University Hospital on August 3, 1964, with chief complaints of genital bleeding and polypoid mass in the vagina.

At the time of admission, she was a well-developed woman and her general condition was normal. Examination revealed grapelike masses arising from the anterior lip of the cervix. Tumor masses had smooth surface and elastic soft

in consistency with occasional area of hemorrhage.

Laboratory findings at the time of admission were as follows : RBC 410× 10^4, Ht. 34.9%, WBC 9200, N. seg. 66.5%, N. band 4.5%, Eosinophils 3.0%, Basophils 1.0%, Lymphocytes 19.5%, Monocytes 5.5%, Hb. 10.5 g/dl, Serum protein 6.8 g/dl, A/G ratio 0.62, Albumin 2.6 g/dl, Globulin 4.2 g/dl, Cholinesterase 0.65 △ph, NPN 20 mg/dl, Urea N 9 mg/dl, Alkaline phosphatase 1.4 u, Cholesterol 52 mg/dl, Phenol turb. test 16.0 u, GPT 2 u. Wasserman reaction of the serum was negative.

Vaginal smear disclosed atypical spindle-shaped cells as shown in Fig. 7. A biopsy from the mass was reported as sarcoma botryoides.

A week later, an abdominal total hysterectomy with bilateral salpingo-oophorectomy was carried out. Examination of the uterus revealed three polypoid masses arised from the anterior lip of the cervix. The largest mass was approximately egg-sized and the smallest one was thumb-sized.

A tumor dose of 4095 r with use of Betatron was administered over a two-month period and anticancerous drug (Mytomycin) was also given. Postoperative course was uncomplicated and she discharged on June 19, 1964. Now she is living, well and free from signs of recurrence of tumor 2 and a half years following operation.

Histopathological Findings :

Tumor was chiefly situated in the submucosal layer and the surface epithelium was preserved in almosy every area (Fig. 8). Tumor was consisted of irregularly arranged, spindle cells with round or oval hyperchromatic nuclei (Fig. 9). The nucleoli were prominent in many of the cells. Mitotic figures were frequent and nuclear pleomorphism was prominent with occasional giant cells (Fig. 10). Though spotted area of necrosis and hemorrhage was seen, myxomatous changes were not so frequent as was observed in Case 1. Fibrillar structure was commonly seen in the cytoplasm of tumor cells, but careful examination of the preparations stained with phosphotangustic acid hematoxylin failed to demonstrate cross striations in the tumor cell cytoplasm (Fig. 11). Reticulum fibers were well developed surrounding individual tumor cells and no alveolar pattern was visible (Fig. 12).

COMMENT

Since its introduction by Pfannenstiel,[6] the term "sarcoma botryoides" has long been applied to uncommon polypoid tumors which arise most commonly in the lower part of the urogenital tract in infancy. In spite of the widely accepted view that the best designation of the neoplasm should be based on the histogenesis or the presumed tissue of origin, the term "sarcoma botryoides",

merely based upon the gross appearance of the tumore, still persists mainly due to the lack of better one.

Many cases are reported to arise in the urogenital area such as the vagina[7-10], the cervix[11], the bladder[12-14] and the prostate[14]. Marcus[11] believed that the müllerian mesenchyme was the site of origin and he suggested the term "müllerian mixed sarcoma" because of the combined presence of fibrous or myxomatous elements, smooth muscles, striated muscles, cartilage, bone or fat. However, similar type of tumors occur not only in the urogenital tract but in the other site of the body. In addition, some cases shows rather monotonous histological appearance and the tumors do not contain combined mesenchymal elements. Therefore, müllerian mixed sarcoma should exclusively applied to the neoplasm which arises from the mesodermal tissue associated with the müllerian duct and furthermore in which varied number of the mesenchymal elements are demonstrable.

In some cases reported as sarcoma botryoides, demonstration of the striated muscle leads to their designation as embryonal rhabdomyosarcoma.[2,5,12,13,14] The designation of embryonal rhabdomyosarcoma is considered improper to some authors[15] on the ground that the rhabdomyoblasts are only a fortuitous circumstance of cellular differentiation not invariably present. In such cases, however, this designation is most preferable since the classification of the neoplasm is made after recognition of the most sufficiently differentiated tissue or cells in the tumor. On the other hand, there are many cases in which the tumor is entirely consisted of undifferentiated mesenchymal tissue, as was presented in this paper. Such immature type is best classified as malignant mesenchymoma. Thus, sarcoma botryoides seems to be an unsatisfactory designation and includes several varieties of malignant neoplasms of mesenchymal origin.

Despite the conflicting opinions as to the histogenesis or the designation of the tumor, sarcoma botryoides has quite characteristic gross appearace in common. It arises from the subepithelial, primitive mesenchymatous tissue of the body cavity that provides the tumors with space for free growth and expansion[8], and the tumors grow in the form of polypoid grapelike masses. Microscopical appearance is rather complicated. In an immature type, the tumor is consisted of spindle-shaped, immature mesenchymal cells with abundant myxomatous stroma. In more matured type, cross striation is occasionally found in the cytoplasm of tumor cells and appearance of multinucleated giant cells is frequent. Occasional examples which contains many mesenchymal tissue and shows characteristics of mixed tumor are also reported[11].

It is commonly accepted that sarcoma botryoides is malignant both histologically and clinically. It has a great tendency to recur locally and makes metastasis sooner or later. Daniel[8] reported 2 of 13 patients with sarcoma botryoides

of the vagina to be living and well without evidence of recurrence, 3 and 5 years after operation respectivelly. According to Stobbe and Dargeon[2], the mean survival time in thirteen patients with this tumor of the head or neck from the onset of the symptoms to death was one year and five months. And two patients were reported to be alive and clinically free from disease for more than 11 years following treatment. Two cases reported in this paper were treated with surgery and preoperative or postoperative irradiation with remarkable effect. Two patients are living and well without evidence of recurrence or metastasis of the tumor up to date. If treatment is to be effective, early diagnosis and immediate radical surgery are necessary. Radiation therapy is also of value as an adjunct to surgery.

SUMMARY

1. Two cases of malignant mesenchymoma (so-called sarcoma botryoides), of which one occurred in the cervix of the uterus and the other arised from the soft palate, are reported. The lesions were treated by a combination of surgery and irradiation with marked effect. Both patients are living and free from recurrence of disease up to date.
2. Brief comment on the histogenesis of the neoplasm has been made and confusion about the designation of the tumors of this type has been pointed out.

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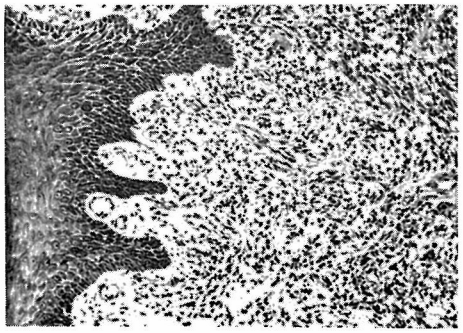

Fig. 1. Tumor tissue is covered by thickened squamous epithelium. Tumor cells are more compact beneath the epithelium than in the center of the polyp. Case 1. H. E. X 100

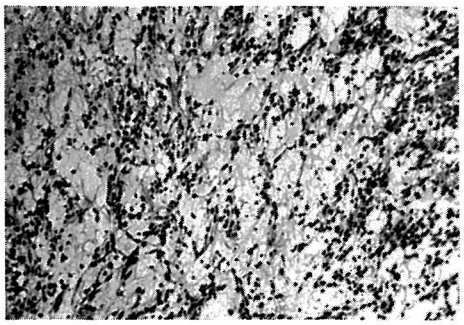

Fig. 2. Very common figure of sarcoma botryoides in which loosely arranged spindle cells are seen in a myxoid stroma. Case 1. H. E. X 100

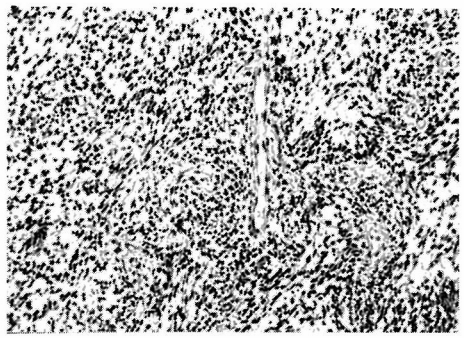

Fig. 3. Tumor is moderately vascular and spindle cells are loosely arranged. Case 1. H. E. X 100

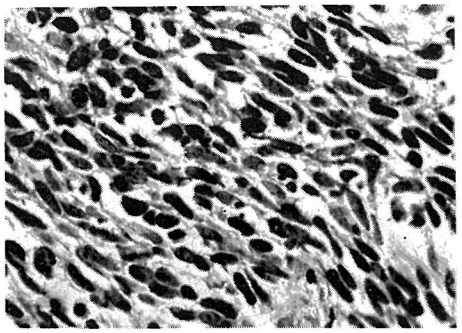

Fig. 4. High magnification of tumor cells which have fusiform, hyperchromatic nuclei with prominent nucleoli. Nuclear pleomorphism is prominent. Case 1. H. E. X 400

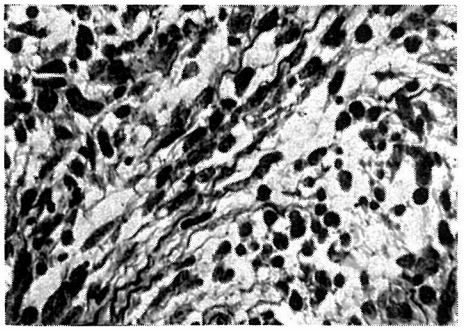

Fig. 5. Tumor cells have elongated, eosinophilic cytoplasm, but cross striation is never observed. Case 1. PTAH. X 400

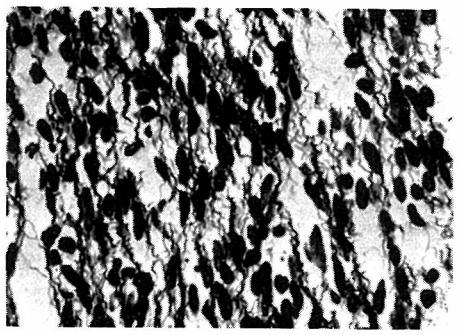

Fig. 6. Reticulum fibers are well developed between individual tumor cells. Case 1. Pap's silver stain. X 400

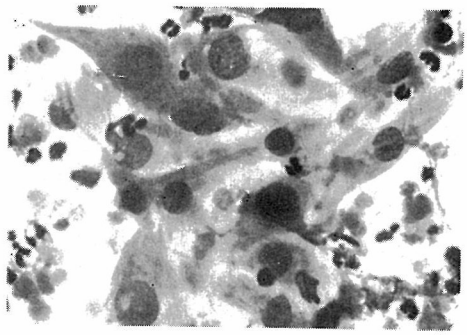

Fig. 7. Tumor cells found in a vaginal smear have fusiform, elongated cytoplasm and hyperchromatic nuclei. Case 2. Papanicolaou stain. X 600

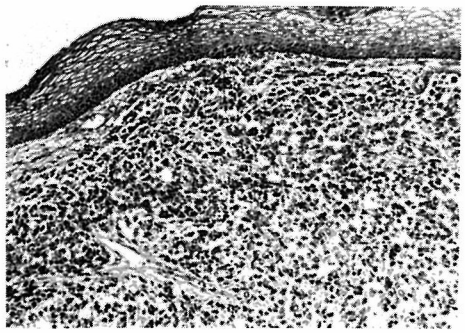

Fig. 8. Polypoid mass, covered by intact epithelium, is composed of haphazardly arranged, sindle-shaped cells. Case 2. H. E. X 100

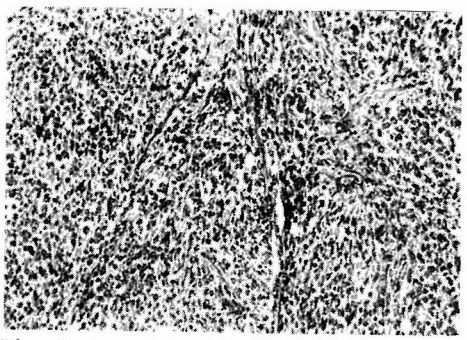

Fig. 9. Around the blood vessels, tumor cells are more compactly arranged, occasionally forming bundles. Case 2. H. E. X 100

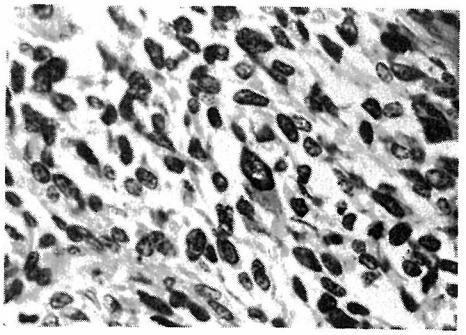

Fig. 10. High magnification of tumor cells which shows prominent nuclear pleomorphism. Case 2. H. E. X 400

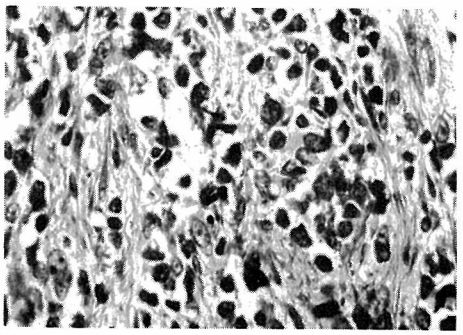

Fig. 11. Fibrillar structure is occasionally seen in the cytoplasm of spindle cells, but it never contains cross striation. Case 2. PTAH. X 400

Fig. 12. Reticulum fibers are intimately bound with the tumor cells and no alveolar pattern is seen. Case 2. Pap's silver stain. X 400