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## Aggressive Angiomyxoma : A Case of Young Female

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Aggressive angiomyxoma of a 7-year-old girl was reported. The primary tumor occurred on the right major labial region as a subcutaneous mass. The extirpated tumor measured 3,0 x 3,0 cm, and revealed a gross appearance of nodular and gelatinous cut surface. Recurrence was noticed 7 months later, on the same location as the primary tumor site. Macroscopic findings of the re-extirpated tumor measuring 2,5 x 2,0 cm was similar to those of the primary tumor.

Histologic features of the primary and the recurrent tumor were virtually identical. Both tumors were composed of two main components i. e., vascular proliferation and fibromyxoid background. Increasing collagenous fibers formed a basic pattern with myxoid stroma, composing a fibromyxoid background. Spindle-shaped tumor cells were sparsely distributed within those fibromyxoid background throughout the tumor lesions. In addition, small vessels of arterial, venous and capillary type appeared randomly or in small groups, in the same fibromyxoid lesions.

The tumor location, specific gross and microscopic features, high recurrency, aggressive nature but no distant metastasis, all these clinicopathologic conditions were considered to support the diagnosis of "aggressive angiomyxoma", which was proposed by Steeper and Rosai<sup>2)</sup> in 1983. Slight variation of the recurrent tumor histology from that of the primary tumor was discussed.

**Key words :** aggressive angiomyxoma

### Introduction

Reports of "aggressive angiomyxoma" (AAM) which was proposed by STEEPER and ROSAI<sup>2)</sup> in 1983 as a new entity of soft tissue tumors have been found rather small in number in literature<sup>2-6)</sup> to this day, especially few in our country.

Recently, we experienced an AAM case of a 7-year-old girl who complained of tumorous swelling in the vulvo-labial region. The tumor recurred 7 months after the first excision. Clinicopathologic examinations were performed for the primary as well as the recurrent tumor, and the specific properties of the present tumors as AAM were confirmed.

### Clinical Course and Gross Pathology

A 7-year-old girl was first admitted to Kumamoto Central Hospital in July, 1989 who complained of the right major labial swelling. According to the parents' explanation on the patient's past clinical conditions, the patient girl had often exhibited behaviors of itchy feeling

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on the vulvo-labial region scrubbing the focal area at desk edges, since 10-month-old baby.

The tumor mass was surgically extirpated. It measured 3,0 x 3,0 cm in dimension, and revealed a gross appearance of nodular and somewhat gelatinous, translucent cut surface, with indistinct border but uniform consistency. Neither necrosis, hemorrhage nor cystic spaces could be found in the tumor parenchyma. Histopathologically, the tumor was suspected of "aggressive angiomyxoma", a new entity of soft tissue tumors.

On follow-up examinations every two months, a recurrent tumor was noticed in February, at the same labial region as the original tumor site. Re-operation was performed for the recurrent tumor on March 28, 1990 with wider excision. The tumor mass existed in areas involving intradermal and subcutaneous tissues, measuring 2,5 x 2,0 cm in dimension. The gross appearance was similar to that of the primary tumor.

### Light Microscopic Findings

Histologic architecture of the primary and the recurrent tumor was virtually identical. Both tumors were composed of two main histologic components i.e., vascular proliferation and fibromyxoid background (Figs. 1 & 2).

Increasing wavy collagenous fibers formed a basic pattern with myxoid stroma described below. However, the density of fibers arrangement varied considerably from area to area, giving an impression of irregular and arbitrary proliferation (Fig. 3). A distinct characteristic was that these fibrous areas accompanied myxoid matrices which were stainable with alcian blue at pH 2,5. These fibrous and myxoid elements appeared rather uniformly in intimate relations throughout the neoplastic areas, composing a fibromyxoid background. Among those fibromyxoid background, spindle cells having uni- or bipolar short cytoplasmic processes were sparsely distributed anywhere in the tumor lesions. These tumor cells possessed ovoid and occasionally cleaved nuclei with fairly dense chromatin and indistinct nucleoli (Fig. 3). Stellate configuration of the tumor cells was not so much frequent as described in literature, in the present case. The cells were bland in appearance. Atypia and pleomorphism were less prominent than those in common sarcoma cells. Mitosis were absent both in the primary and the recurrent tumors.

High vascularity composed another distinction of the tumors. Small vessels of arterial, venous and capillary types were distributed randomly or in small groups within the fibromyxoid

### Explanation of Figures

All the photomicrographs (Figs. 1-4) were taken from the primary tumor. Tissues were stained with hematoxylin and eosin.

Fig. 1 : A thick-walled arterial vessel can be seen in the center of the micrograph with prominent smooth muscle hypertrophy and adventitial fibrosis, resulting in the vascular channel being narrowed. Adjacent to it, another vessel (probably venous) is visible in congestive fashion.

Fig. 2 : Several small vessels exist in the fibromyxoid background. Two capillary type vessels dilate markedly, containing organizing thrombi in their lumina.

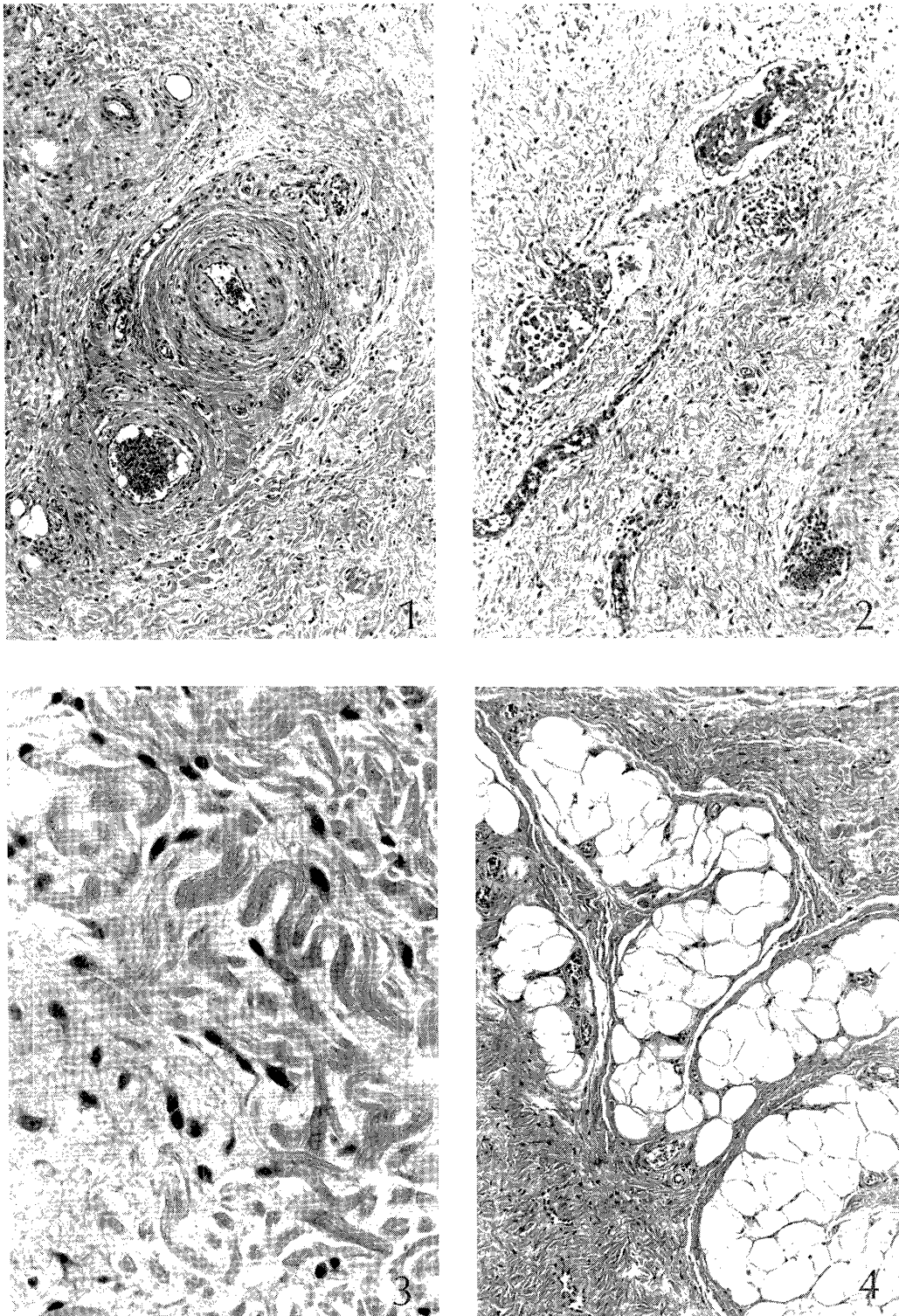


Fig. 3 : High power photomicrograph of a fibromyxoid background. Arrangement of wavy collagenous fibers look denser in the upper right, but less dense in the lower left field. Spindle-shaped tumor cells are distributed within the fibromyxoid background.

Fig. 4 : Bordering area of the tumor with subcutaneous fat tissues. The fibromyxoid proliferation is invasively spreading downward, dividing the fat lobules into more irregular groups.

lesions (Figs. 1 & 2). Some capillary type vessels dilated markedly with capillary sinus-like appearances, containing organizing thrombi in their lumina on occasion (Fig. 2). Although venous vessels dilated also in congestive fashion, most arterial vessels appeared in thick-walled features because of hypertrophic muscle layers and adventitial fibrosis, resulting in the vascular channels being narrowed (Fig. 1).

Small groups of fat cells and peripheral nerve bundles were found entrapped haphazardly within the tumor parenchyma. These well-differentiated tissue structures did not seem to be part of the true tumor components, but they seemed to be an accidental participation of normal tissues in the pathologic lesions.

The tumor margins were indistinctly adjacent to the surrounding normal tissues. In upper side of the tumors, the fibromyxoid tissues looked to be gradually disappearing into the dermal collagen. In the basal portions, however, the fibromyxoid proliferation was invasively spreading downward, dividing the subcutaneous fat lobules into more irregular groups (Fig. 4). No sharp demarcation was detectable in any bordering areas of the primary as well as the recurrent tumor.

Immunohistochemical stains of the tumor tissues were tested for desmin, vimentin, S-100 protein and factor VIII. The spindle-shaped tumor cells stained positive only for vimentin and negative for desmin, S-100 protein and factor VIII.

### Discussion

In 1983, STEEPER and ROSAI<sup>2)</sup> proposed a new disease entity "aggressive angiomyxoma" for a myxoid tumor group which occurred preferentially in perineal, vulvo-vaginal or pelvic regions of females, presenting nine cases they collected and a short note of four additional female cases they experienced subsequently. Two years later, in 1985, BEGIN et al<sup>3)</sup> reported other nine cases of aggressive angiomyxoma (AAM) including two males, with an evaluation for the descriptions by STEEPER and ROSAI, and added some new knowledges for the tumor.

In diagnosis of our case presented here, attention was given to the patient's clinical and clinicopathologic conditions, gross and histopathologic characteristics of the extirpated tumors, which were mostly consistent with those described in above authors' reports. Age and sex, tumor location, gross appearance, infiltrative nature but no distant metastasis, high recurrency, special histologic features, all these conditions of the present case would be able to support fully the diagnosis of "aggressive angiomyxoma".

We suppose, however, that the histologic details of AAM may be variable within some limits in individual cases, or even in the same patient, the microscopic features between primary and recurrent tumor may vary by unknown influential factors. In our own case, the general histologic features of the primary and the recurrent tumor were basically identical. While in details, vascularity of the recurrent tumor decreased as compared with that of the primary tumor. As the result, the fibromyxoid constituents predominated in the lesions, giving a fibroma-like or scar-like impression. Thus, the recurrent tumor seemed to have appeared in some variation from original AAM histology. Before the present case reporting here, OHTSUKA<sup>5)</sup> made a short report, in 1987, of a 38-year-old female who had been bearing a major labial tumor, under the probable diagnosis of AAM for the excised mass. Although the diagnosis was confined to suspicion because of lacking in definitive histologic characteristics,

we still suppose that case to be a variant of AAM, if the histologic features of AAM involve some variety range.

In addition to the fibromyxoid features of AAM, we regard that aberrant and high vascularity is another important characteristic. In our own case, not only the vascular components showed random distribution, but they were composed of arterial, venous and capillary type vessels intermixedly. Besides, respective type vessels appeared mostly in abnormal fashions, such as thick-walled arteries, congestive veins, and sinus-like dilated capillaries. We understand that all these vascular abnormalities exhibit profiles of angiomatous and neoplastic proliferation.

As for ages of occurrence of AAM, it has become evident that AAM involves wide age spectrum by reviewal of the cases reported so far. According to the summarized data by KAWAI et al.<sup>6)</sup> in 1988, the ages ranged from 18 to 63-year-old including two males. Our present case, therefore, extended the AAM younger age spectrum up to 7-year-old.

With respect to differential diagnosis of AAM from other myxoid tumors benign and malignant, STEEPER and ROSAI<sup>2)</sup> discussed in their article various tumors liable to lead misdiagnosis. Two years before above authors' report, there could be found a general description by MACKENZIE on "The myxoid tumors of somatic soft tissues" in 1981. However, we don't think we need to consider such various tumors which can be separated easily from AAM, for diagnosis of our present tumor. Yet, it would be necessary to take several myxoid and fibroid tumors into consideration. Because, the present tumor formed its basic texture on prominent collagenous and fibromyxoid proliferation. In diagnosis of our present tumor, the following myxoid tumors will be excluded from AAM entity by their characteristics mentioned below. Myxoma; benign non-aggressive nature, low vascularity, low recurrence rate. Myxoid MFH; preferential ages (late adulthood) and locations (extremities), storiform arrangement of fibers. Myxoid liposarcoma; various features of lipoblast including spider web and signet ring cell type. Myxoid neurofibroma; probably the most similar histology to AAM. admixture of collagen and finer neurogenic fibrils stainable with silver impregnation, electron microscopic features of Schwann cells, low vascularity.

Regarding the biological nature of the spindle and stellate tumor cells, STEEPER and ROSAI, BEGIN and associates were of different opinions. STEEPER and ROSAI postulated myofibroblastic nature of those cells, standing on the immunohistochemical and electronmicroscopic findings. BEGIN and associates, on the other hand, regarded the cells as fibroblastic nature and differentiation. Unfortunately, we have little informations in this problem because of lack of an electron microscopic study of the present case, although the immunohistochemical results are indicating fibroblastic differentiation of the tumor cells.

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