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혈관육종 환자에서 두피의 아전절제술

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Near Total Excision in Patients with Angiosarcoma on Scalp

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Purpose: Angiosarcoma is a rare malignant neoplasm of endothelial type cells that line vessel walls. It tends to occur in aged male and the prognosis of angiosarcoma is very poor because of frequent local recurrence and early metastasis. The treatment regimen is yet to be established from its rare occurrence but the wide excision in early stage is known to be the most effective. The authors report two cases of near totally excised angiosarcoma with more than a safety margin of 5 cm.

Methods: The two subjects were aged male patients, one of the two was diagnosed with angiosarcoma from our institution confirmed by the biopsy. The other one went through the wide excision with a safety margin of 2 cm and split-thickness skin graft but local recurrence was observed. The two patients underwent near total excision with more than a safety margin of 5 cm, leaving only the periosteum. After confirming that the angiosarcoma had not infiltrated the excision margin, reconstruction with split-thickness skin graft was performed.

Results: Based on 6 months and 24 months post-surgery assessment, no local recurrence or remote metastasis in the lungs, liver, bones, and lymph nodes at the neck, where remote metastasis is common, was reported by the two subjects who underwent near total excision with a safety margin of 5 cm.

Conclusion: Angiosarcoma has very poor prognosis from its frequent recurrence and metastasis. To enhance the survival rate of angiosarcoma patients, early diagnosis, timely surgical treatment, and radiotherapy after surgery are critical. In addition, authors suggest that it is necessary to further study the efficacy of wide excision using a wider safety margin as much as possible, and to apply this to more cases.

Key Words: Angiosarcoma, Scalp, Total excision

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I. INTRODUCTION

Angiosarcoma is a rare malignant tumor originating from the endothelial cells that account for approximately less than 1% of all soft tissue tumors.[1] About half of all angiosarcoma cases occur in the head and neck area, but it is especially common in the scalp. It has been known to occur in males in their sixties to seventies.[2] Generally, its five-year survival rate is about 15%, which is a very poor prognosis among soft tissue tumors, and wide excision in the early stage is the best treatment option.[1,3] It is hard, however, to decide the infiltration range and the surgical margin because the clinical boundary of its lesion is vague, it is commonly accompanied by inflammatory erythema near the lesion, and it has the characteristics of relatively rapid local and remote metastasis besides discontinuous infiltration, such as satellite lesions.[4] These authors formerly performed wide excision with a safety margin of 3 cm for two cases of angiosarcoma on the scalp, both of which had poor prognosis. In this article, authors report another two cases which are under follow-up monitoring after near totally excised angiosarcoma with a safety margin of 5 cm.

II. IDEAS AND INNOVATIONS

Case 1

A 71-year-old male who had a dark-blue papule accompanied by pruritus on the bregma underwent radiotherapy after wide excision with a safety margin of 2 cm and split-thickness skin graft, after being diagnosed with angiosarcoma in another hospital. Multiple dark-blue papules less than 1 × 1

cm in size, however, and an erythematous lesion, occurred near the previous surgical site, which the patient presented to these authors' hospital. The biopsy showed manifestations of the recurrence of angiosarcoma, but no suspicious manifestations of metastasis were found in the radiological study.

The blood vessels, which were covered with various-shaped atypical tumor cells on the hematoxylin and eosin stain over the whole dermis, underwent anastomosis, as shown in the histopathological tests. Mitotic figures were often detected, and there were diffuse infiltrations to subcutaneous fat, with proliferated fusiform vascular cells in some parts. Moreover, positive to factor-VIII-related antigens were shown in the immunohistochemistry test.

Near total excision with a safety margin of 5 cm was performed near the tumor site, leaving only the periosteum. After confirming that the angiosarcoma had not infiltrated the basal layer and its surroundings in the frozen section during the surgical procedure, reconstruction with split-thickness skin graft was performed. Two weeks after the surgery, the thread was removed, and one month after the procedure, radiotherapy was performed. There was no suspicious manifestation of recurrence upon outpatient follow-up for 24 months after the surgery. In addition, no metastasis manifestation was observed in the lungs, liver, bones, and lymph nodes at the neck, where remote metastasis is common (Fig. 1).

Case 2

A 71-year-old male patient visited these authors' hospital with a lesion at the right parietal scalp, which had emerged two months earlier. A 3.5 × 3.5 cm ulcerative lesion and an erythematous lesion with obscure boundaries were observed in the physical examination, without any significant finding. Via biopsy, the patient was diagnosed with angiosarcoma, with the same pathological outcomes as in the previous cases. In the radiological test, no finding was obtained that would

Fig. 1. Preoperative view of the 71-year-old male patient with angiosarcoma. Local recurrence of an angiosarcoma was found two months after the initial wide excision with a safety margin of 2 cm. (Left) Variable size of dark brownish and erythematous lesions at the surgical site. (Center) Near total excision with over safety margin of 5 cm. (Right) Postoperative view after one month.

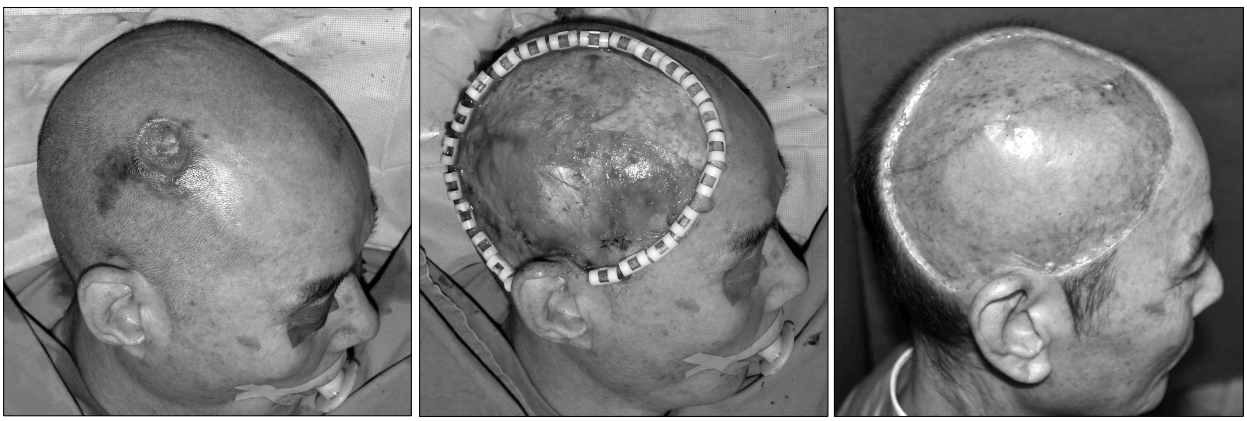

Fig. 2. (Left) Preoperative view of the 71-year-old male patient with angiosarcoma. 3.5 × 3.5 cm sized lesion on the parietal scalp with ill-defined erythematous lesions. (Center) Wide excision with over safety margin of 5 cm. (Right) Postoperative view after two month.

lead to a suspicion of metastasis. Like the previous patient, this patient underwent wide excision with a safety margin of 5 cm near the tumor site, the only remaining periosteum. After confirming that there was no angiosarcoma infiltration in the boundaries and basal layer of the frozen section, it was reconstructed via split-thickness skin graft. The suture was removed in two weeks, and radiotherapy was initiated one month after the surgery. No finding that could lead to a suspicion of local recurrence or remote metastasis was reported six months after the surgery, in the outpatient follow-up (Fig. 2).

III. DISCUSSION

Angiosarcoma, which originates from the endothelial cells, is a rare tumor accounting for less than 1% of soft tissue tumors.[1] Besides the skin, it can be developed in the vascular endothelial cells, including the breast, liver, skeletal, and striated muscles. It can be classified into three different types: angiosarcoma occurring on the scalp and face of the aged, secondary angiosarcoma in chronic lymphedema, and angiosarcoma occurring after massive radiation therapy. About 50% of angiosarcoma cases begin in the head and neck area, but angiosarcoma is especially common in the scalp. It occurs mainly in people in their sixties to seventies, and has a known tendency to occur mainly in males, with a 3 : 1 male : female ratio.[2]

Angiosarcoma generally starts from a bluish spot, but the observed clinical manifestations vary from asymptomatic cases to cases with a necrosis crust accompanied by bleeding.[1,2] It requires differential diagnosis from inflammatory diseases such as seborrheic dermatitis, cellulitis, erysipelas, and bruise due to its pattern of non-specific and gradual dispersion at the initial stage, and from cancers such as hemangioma, lymphosarcoma, malignant melanoma, and squamous-cell carcinoma after its development. Moreover, definite diagnosis is delayed in the case of non-specific symptoms because in such cases, only conservative treatment is performed, without biopsy. The physicians' ignorance due to the low incidence rate of angiosarcoma is another reason for delayed diagnosis. Considering the rapid metastasis pattern of angiosarcoma, a delay in wide excision due to late diagnosis may seriously affect the prognosis. Therefore, if a case shows clinical patterns similar to those of angiosarcoma, or is suspicious for a differential disease, it is crucial to correctly diagnose it earlier through biopsy.[1] Wide excision with additional radiotherapy is currently the best option for early stage patients when excision is possible, but it has been known that advanced angiosarcoma patients who experienced recurrence in spite of this treatment showed poor prognosis after radiotherapy and chemotherapy.[1,5] In addition, the tumor size and the surgical excision of the initial lesion are important prognosis factors, and poor prognosis factors have been reported with tumor sizes over 5 cm, metastasis cases, undifferentiated tumor cells, old age, and infiltration up to the surgical-site margin.[4,6]

The pathophysiological patterns of angiosarcoma are simultaneous infiltrations up to the dermis and subcutaneous fat, and various differentiation levels even in the same lesion, that is, less differentiated in the peripheral areas and well differentiated in the central region, forming a severely infiltrated nodal ulcer consisting mainly of single-layered endothelial cells. The vessels that form endothelial cells are irregularly connected to one another.

Less differentiated blood vessels consist of atypical, cuboid endothelial cells, form crooked sinuses that significantly expand, and make a papillary projection inward with proliferated endothelial cells. Much less

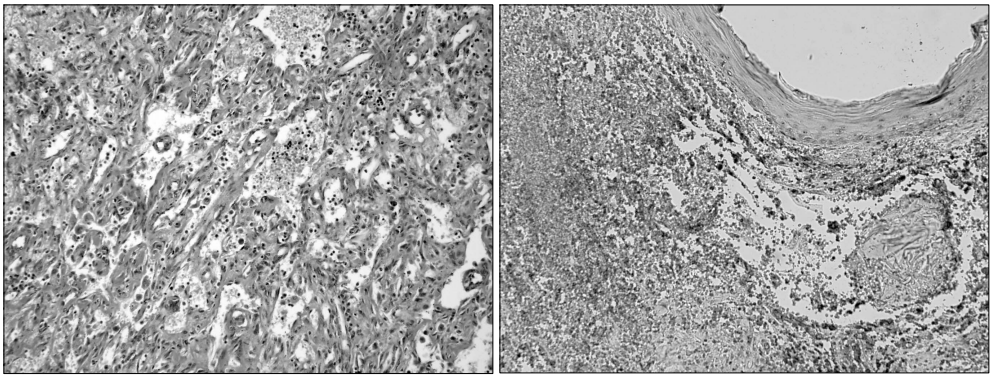

Fig. 3. Histological findings of angiosarcoma. (Left) Dilated vascular spaces lined by plump, atypical endothelial cells dissecting through collagen fiber (H & E stain × 200). (Right) Immunohistochemical stain for CD31 shows focal immunoreactivity (× 200).

differentiated parts show weak vessel development and are infiltrated by spindle cells, like cords. Considering these histological and clinical patterns, approximately 75% of angiosarcoma cases show a mainly moderate or higher level of differentiation in the macule and plaque, while the nodosity shows less differentiation.[7] Immunohistochemistry is helpful for the diagnosis and differential diagnosis of angiosarcoma, but angiosarcoma is shown to be positive for CD31 and CD34, which have high specificity to hematologic tumors, and are often positive for factor-VIII-related antigens and Ulex europaeus 1 lectin (Fig. 3). About 35% of angiosarcoma cases are shown to be positive for cytokeratin and negative for EMA and S-100. Whether angiosarcoma originates from the epithelial cells of the blood vessels or from the lymphatic vessels is controversial, but the existence of RBC in the blood vessels, and the expression of CD31 and factor-VIII-related antigens, show a likelihood of originating from the endothelial cells of the blood vessels.[8]

As angiosarcoma has been known to be extraordinarily malignant among soft tissue tumors, wide excision is the best option in the early stage.[1,2] Moreover, it has been reported that many cases had already been significantly developed at the time of detection, and that the limited feasibility of wide excision was related to local recurrence, remote metastasis, and the subsequent decrease of the survival rate.[1] Furthermore, radiotherapy or chemotherapy is being carried out, but the treatment has not been well established. Wide excision has been reported to be the most useful option for the basic treatment principle, but it is difficult to decide the surgical margin for such.

Previously, these authors experienced two patients who expired after undergoing surgery with a safety margin of 3 cm, due to the complications caused by the metastases to the liver and the lung. Although continuous follow-up monitoring is required, no finding of local recurrence or remote metastasis has yet been observed in the two patients who underwent wide excision with more than a safety margin of 5 cm. Of course, the other factors that showed poor prognosis should be considered in the cases where a safety margin of 3 cm was applied, such as the partial removal of the outer table of the skull even though brain metastasis had not occurred, the fact that the lesion was somewhat large, and the fact that there was not a single lesion. It is meaningful, however, that local recurrence or remote metastasis was not observed in the first case, where wide excision with more than a safety margin of 5 cm was applied, in spite of the poor prognosis of local recurrence after the surgery. Therefore, early diagnosis, a timely surgical procedure, and radiotherapy after the procedure are very important to improve the prognosis in patients with angiosarcoma. Moreover, further studies applying this to more cases are required to understand the relationship between wide excision using a wider safety margin and the survival rate.

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