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Case Report

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Intracranial Invation From Recurrent Angiosarcoma of The Scalp

Angiosarcoma of the brain, either primary or metastatic is extremely rare. Moreover, angiosarcoma metastazing to the brain is also highly unlike to occur when comparing with metastases to the other organs. Thus, an ideal treatment strategy has not been established. A 67-year-old man with past surgical history of a scalp angiosarcoma underwent surgical resection of intracranial invasion. Because of wide scalp flap excision and resultant poor vascularity of the scalp flap, additional radiation was not provided. Because adjuvant therapy is impossible due to poor scalp condition, more careful but ample resection of the primary lesion is essential to conduct initial operation.

KEY WORDS : Brain neoplasm · Metastasis · Scalp Angiosarcoma · Scalp flap .

INTRODUCTION

Angiosarcoma is an infrequent malignant vascular neoplasm originating in the face, scalp, liver, skin, and soft tissues elsewhere in the body[10]. Its involvement of the brain, either primary or metastatic, is an extremely rare phenomenon with very few descriptions available in the literatures[1-3,7,9,11-13]. When we consider its aggressive biological behavior, such as local invasiveness, high recurrence rate and proximity to the adjacent overlying scalp, its rare incidence is above our apprehension. There seems to be a naturally privileged barrier hampering local spread and con-sequent seeding in the cranial cavity.

We describe a case of primary angiosarcoma of the scalp that subsequently invaded to the brain leading to the intraparenchymal hemorrhage. With this case presentation, we emphasize adequate scalp management in the initial surgical management.

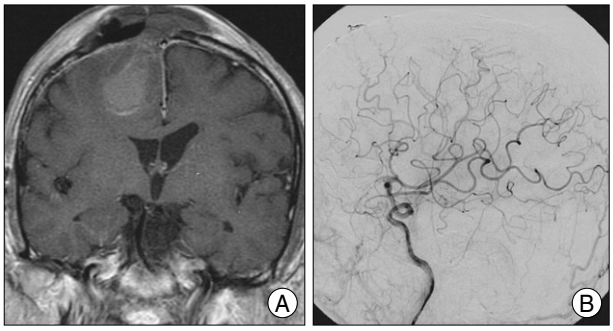

Fig. 1. A : Enhanced T1-weighted magnetic resonance image showing subacute hemorrhage on the frontal motor cortex with perilesional edema. The hemorrhage is contiguous with overlying skull and scalp. B : Transfemoral carotid angiogram shows no tumor staining.

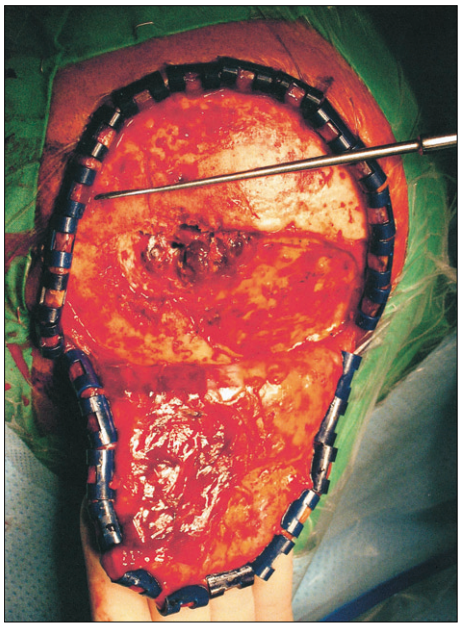

Fig. 2. Intraoperative photograph shows dark reddish hematoma in the scalp, skull and brain.

CASE REPORT

A 67-year-old man presented with left-sided motor weakness for 4 days. He was alert and oriented, free of any cranial neuropathy, but had left hemiparesis (Grade III/IV). He had undergone surgical resection of angiosarcoma of the scalp and skull vault one

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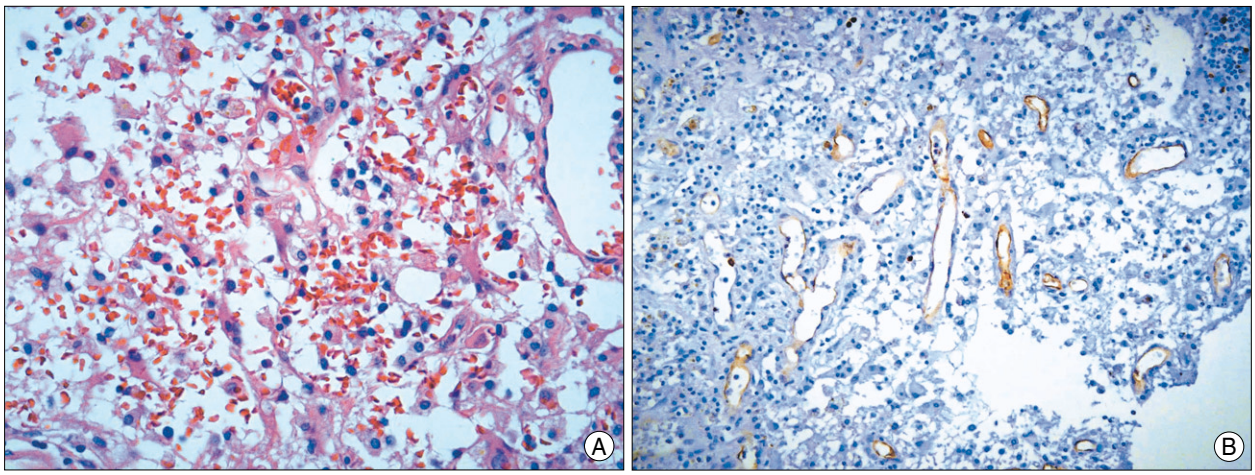

Fig. 3. A : Histopathological examination shows epithelioid cells with necrosis, vascular channels and anastomosing network of sinusoids (H&E, x400). B : Immunohistochemical staining for CD31 shows strong positivity (IHC, x400).

year prior to admission. Also, reconstruction with skin free flap and artificial bone cement was performed simultaneously. At this time, magnetic resonance (MR) imaging scan revealed minimally enhancing subacute hemorrhage in the frontal motor area with perilesional edema. The hematoma was contiguous with overlying skull and scalp (Fig. 1A). Carotid angiogram shows no specific finding (Fig. 1B). Surgical resection was performed to all involved structures. They were simultaneously reconstructed with resin (polymethyl methacrylate) and lyophylized dura. During the operative procedure, semisolid dark purple hematoma was evacuated (Fig. 2). Pathologic examination verified the presence of epithelioid cells with necrosis, vascular channels, and anastomosing network of sinusoids as well as positive staining for vimentin, CD34 and CD31. The final diagnosis of cerebral angiosarcoma invaded from the scalp was made on the histologic report (Fig. 3).

Postoperatively, neither adjuvant chemotherapy nor radiotherapy were provided because of poor scalp condition, that was thinned by trimming subcutaneous tissues and patched in some area. The patient's postoperative course was uneventful and head computed tomograpy (CT) scan revealed no remarkable enhancing lesion.

Two months later, it was noted that his scalp was bulged, and pulsatile. Moreover, necrotic change was strongly suspected (Fig. 4A). MR image revealed a small wedge-shaped enhancing lesion in the parasagittal area adjacent to the previous operative field at the coronal scan (Fig. 4B). Radical resection and rotational scalp flap were successfully conducted with unremarkable intradural exploration (Fig. 5). Histologic diagnosis of angiosarcoma of the scalp was made again and MR imaging at postoperative 2 weeks revealed no definitive evidence of enhancing lesion. He showed obscure postoperative recovery, eventually we lost the chance to follow-up him because of denial to further surveillance.

DISCUSSION

Primary intracranial sarcomas are uncommon and are reported to comprise 1 to 2% of all primary intracranial neoplasms[1]. Of these tumors, angiosarcoma is particularly rare, accounting for less than 1% of all sarcomas[1,10,11]. Although it may occur in the face, scalp, liver, trunk, heart, extremities, and spleen, the occurrence of primary cerebral angiosarcomas is quite anecdotal. In addition,

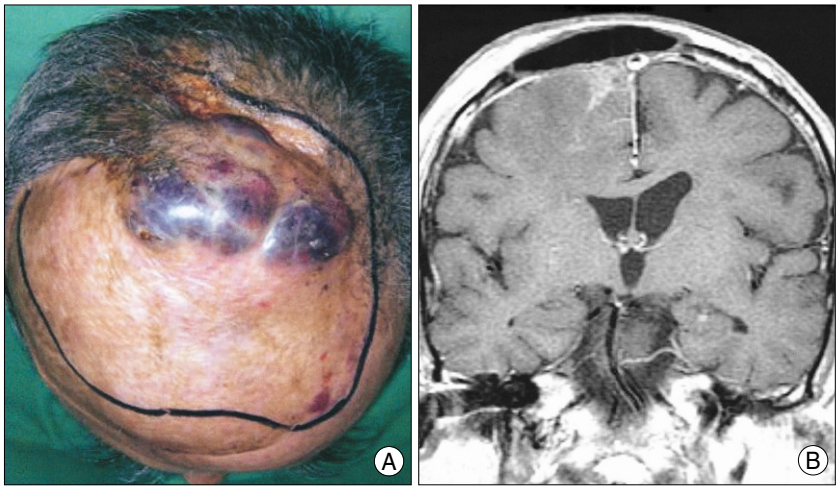

Fig. 4. A : Photograph shows scalp bulging with accompanying necrosis in the previous operative area. B : Enhanced coronal T1-weighted magnetic resonance image shows slightly enhancing lesion on the parasagittal area adjacent to the previous operative field.

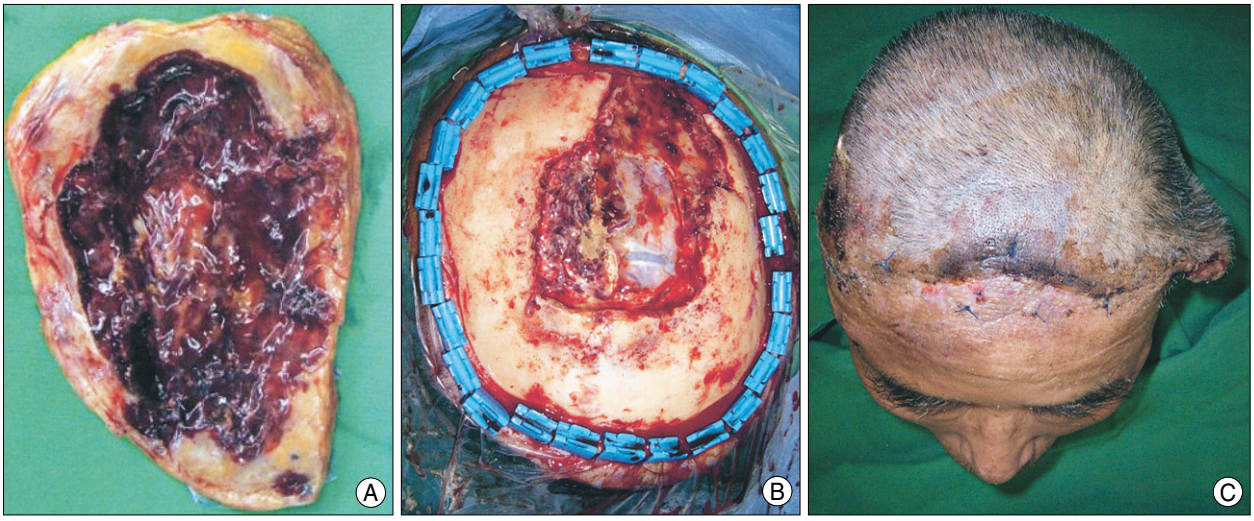

Fig. 5. A, B : Intraoperative photographs show tumor cell invasion in the scalp and dura adjacent to previous artificial bone flap. C : Postoperative photograph shows reconstructive lesion after rotational scalp flap.

angiosarcoma metastasizing to the brain is also highly unlike to occur when comparing with metastases to the other organs such as lungs, bones, liver, lymph nodes, adrenal glands, spleen, and pleuras[7,10].

Only a few cases of cerebral metastasis from angiosarcoma of the heart have been described in the literature, with particular relevance to intracerebral hemorrhage[4].

Irrespective of specific cerebral location, we suggest that immediate surgery is mandatory to prevent lethal evolution as a result of frequent rebleeding and aggressive local recurrence. Thus, the initial approach to patients with cerebral angiosarcomas should be direct surgical removal. Survival of patients with angiosarcoma is generally poor with propensity for both local recurrences and distant metastases[1]. Because the extensive microscopic spreads are common with this disease, it needs to be resected circumferentially by dissecting around the tumor rather than intratumoral debulking.

The role of adjuvant treatment on the survival of patients with cerebral angiosarcoma is not well defined because of the rarity of the disease. Several authors report that radiation therapy has been offered as a possible adjuvant therapy[5,6,8]. Small lesions seem to be treated either by surgery or radiotherapy. Possible lower incidence of metastases in patients provided by radiotherapy may indicate that the treatment of choice is radiotherapy alone or radiotherapy to a wide field following radical excision of the tumor.

As stated above, both skull and dura mater play as a natural barrier to prevent direct invasion of the angiosarcoma to the brain from the overlying scalp. For this reason, a small gap between skull and artificial resin, that created from the first resection, seemed to have provided a conduit for contagious spread from the remaining scalp neoplasm. In the present case, the patient had thin skin less than 1/3 of the original thickness and resultant poor vascular supply. Moreover, repeated local recurrences of the scalp expanded progressively near the initial operative field. For this reason, we could not perform postoperative adjuvant therapy and it was inevitable that we strived to manage the scalp wound problem only.

CONCLUSION

This unusual case highlights the need to consider direct invasion of the scalp angiosarcoma with highly aggressive biological behavior, such as local invasiveness and high recurrence rate, in circumstances under the collapsed natural barrier.

Surgical resection with wide but careful negative margin is the gold standard treatment, because there exists controversy for the efficacy of postoperative adjuvant therapy or localized problem of scalp wound.

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