Case Report

Giant Scalp Angiosarcoma With Intracranial Extension: Case Report

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Abstract

Angiosarcoma is a rare and extremely aggressive soft tissue tumour which generally occurs in head and neck region, particularly in elderly people. They may arise as small plaques or multifocal nodules in the scalp. The diagnosis is not easy before arisen of neurological symptoms and pronounced enlargement in the scalp. Treatment is closely related to the course of the disease. Surgical treatment consists of wide resections and reconstruction. Radiotherapy and chemotherapy may be indicated in cases of recurrent large lesions, focal or distant metastases. This study presents a 78-year-old female patient with a noticeable swelling on the left side of her head and history of bleeding in the scalp. Mass lesion with intracranial invasion was removed with wide surgical margins and reconstruction followed by adjuvant radiotherapy.

Keywords: Angiosarcoma, Scalp, Treatment

INTRODUCTION

Angiosarcoma is an extremely malignant vascular tumour of endothelial origin that generally locates in the head, face and neck regions\(^{(19,26)}\). It forms up approximately 10% of all the sarcomas that develop in the head and neck region. Angiosarcoma is rarely seen in the brain as a primary or metastatic tumour. Older population is much more affected\(^{(6,12)}\). Prevalence is higher in the Caucasian males. Chronic lymphedema and radiation exposure are suspected as predisposing risk factors in some of the patients. The disease may develop as single or multiple, nodular,
bleeding or ulcerated plaque\textsuperscript{(17,21)}. Metastasis may develop in regional lymph nodes or lung parenchyma\textsuperscript{(16)}. Diagnoses may prove difficult until these tumours become symptomatic and large. This type forms up approximately 1\% of all the sarcomas and less than 0.1\% of head and neck region tumours which results in late diagnosis. The invasion and recurrence rates are high\textsuperscript{(1,24)}. The treatment options of these tumours generally include wide resection and reconstruction. There is a greater risk of recurrence if not radically removed. Radiotherapy or chemotherapy may be applied to the tumours which are not totally removed, and which are extremely large and which present with distant metastasis\textsuperscript{(10,13)}. Nevertheless, the prognosis of these patients is poor and is not related to the sex, age, tumour localization and clinic. The size of the tumour is associated with recurrence and prognosis which leads us to the conclusion that, early diagnosis and radical surgery are of significance however survival rate among these cases is less than 30\%\textsuperscript{(3,19,26)}.

**CASE PRESENTATION**

A 78-year-old female patient was admitted to our clinic with the chief complaints of seizure and rapidly growing bulging on the left side of her head. Physical examination revealed a mass lesion measuring approximately 10 cm with irregular margins and bleeding points on the left frontoparietal region. Neurological examination was uneventful. No distant metastasis and/or lymph nodes involvement were determined in systemic examinations. Cranial MRI showed a hypervascular tumoural lesion measuring approximately 8 – 10 cm which is fed from the branches of external carotid arteries on the left frontoparietal region. Left superficial temporal artery (STA) was larger when compared to the right side artery. Many arterial vascular structures were seen in the tumour (Fig. 1). Cerebral angiography revealed many vascular structures which were considered to arise from the meningeal artery on the left frontotemporal region. The vascular branching was determined to enhance in the left middle cerebral artery trifurcation level (Fig. 2). The patient was operated as a result of the radiological and physical treatment findings. Tumour was totally removed. Reconstruction was performed with scalp flap transfer. No complications were observed postoperatively. Control MRI did not show any recurrence (Fig. 3). The pathological diagnosis of the patient was considered to be angiosarcoma. Many atypical mesenchymal cells were noted and some of these cells consist of many cystic dilated vessels while rest of cells was presented with pronounced fibrotic stroma in some sites. These cells were seen in the holes of vessel structures and cleft like formations. Immunohistochemical examination exhibited tumour cells that were stained positively with factor 8, CD31, vimentin, LMW – CK. Glycogen was positively stained with PAS diastase (Fig. 4). Patient received postoperative radiotherapy. Follow-up period was uneventful in the subsequent two years but in the second year control MRI, two small solid masses on the left parietal dural surface and recurrent lesions in two different regions of the left frontoparietal bone were noted. The patient with normal neurological course was referred to the medical oncology department for surveillance.
**Fig 1:** Axial T1 weighted MR image demonstrating a large, hyperintense scalp lesion that is extending to the calvarium and brain parenchyma of the left temporoparietal region. Tumour mass feeding from approximately 8 – 10 cm External Carotid Artery in the level of calvarial bone on the left frontoparietal was seen as a result of T1 weighted axial MRI. Many arterial vascular structures were seen inside the tumour.

**Fig 2:** Cerebral angiography showing a large, hypervascular tumoural lesion originating from the left meningeal artery. Note that many vascular structures are encircled by the tumour.

**Fig 3:** Gross total tumour excision is seen on control T1 weighted axial MR image.

**Fig 4:** Histopathological specimen shows epithelioid cells with necrosis, vascular channels and anastomosing network of sinusoids (H&E, ×400).
DISCUSSION

Angiosarcoma is an uncommon soft tissue tumour which originates from endothelial cells\(^{17,25}\). Primary intracranial sarcomas are rather uncommon and they form up 1 – 2% of all intracranial tumours. Angiosarcomas form up less than 1% of all sarcomas. They may develop in liver, heart, spleen, extremities, face and scalp\(^{21,25}\). They may be metastatic in the brain, as well. They may also be metastatic in lung, bone, liver, lymph glands, adrenal glands, spleen and pleura\(^{13}\). Intracranial haemorrhage may accompany with brain metastasis\(^{8}\). On the other hand, cutaneous angiosarcomas such as scalp involvement may develop in patients who are treated for chronic lymphedema, radiotherapy, breast cancer and renal transplantation\(^{7,16}\). The theories suggesting that sunlight exposure and actinic keratosis cause that disease were refuted with the cases that are seen in the patients with protected hair scalp\(^{18,20}\). Kasabach – Merritt syndrome associated diffused angiosarcoma cases were reported in the literature\(^{22}\). Some of the factors, despite being predisposing, should be evaluated. Some of these factors include the history of herpes zoster, telangiectating nevus, vascular and lymphatic pathologies, arteriovenous fistula, chronic osteomyelitis, thorotrast and polyvinyl chloride reaction. Most of the patients develop scar like macule, or nodule as well as, endured erythematic nodules, mushroom shaped masses, ulcerated or bleeding lesions\(^{5,9,20}\). Generally males between 60 – 75 years present with scalp angiosarcoma. The prevalence ratio in males and females is 1:2. The survival rates of these tumours with the high tendency of recurrence and distant metastasis show poor survival rates. The lymph node (LN) metastasis rates of the tumour are superior to all other soft tissue tumours located at the head and neck region. Moreover, the metastasis rate is over 50% and frequently develops in lungs and livers\(^{15,16}\). The high tendency to metastasis is associated with the absence of vascular endothelial cadherine (VE – cadherine), as the vascular endothelial cadherine is seen in the normal endothelium margins. Low grade angiosarcomas are well differentiated while the high grade lesions are poorly differentiated. The pleomorphic cells and bleeding malformed structural sites include the cells with pleomorphic nucleus and pronounced mitotic activity\(^{2,5}\). Both forms are characterized by local growth. There is no exact correlation between stage of tumour and survival\(^{18}\). As they are locally aggressive and show high recurrence rates, in the patients with scalp and cranial involvement radical surgery is necessary\(^{9}\). Recurrence rates in the cases with subtotal resection are higher. Multicentric characteristics and microscopic level invasion may aggravate the angiosarcoma treatment on the scalp\(^{15}\). The treatment option until the margin without tumour is histological large excision. This situation is directly associated with prognosis. Due to large excision, primary wound closure is not possible most of the times\(^{10}\). Reconstruction may be performed with the alternatives such as split – thickness skin grafts, local flaps, and free flaps\(^{4,18}\). Reconstruction is of importance in order to prevent the scalp necrosis and infection particularly in the older patients. Except the patients receiving radiotherapy in advance and with cranium defect, skin grafts are most frequently employed. Local rotation flaps are indicated if the pericranium is removed and the defect is not so large. In order to prevent the recurrence in case of large angiosarcomas, scalp may largely and totally be removed. In that sense, large defects are reconstructed with free flaps\(^{4,11}\). Small doses of radiotherapy may be applied to the patients postoperatively with diffused multifocal lesions. However, it has been reported that radiotherapy is not effective...
on survival rate in the patients with radical tumour remove. Large doses of brachytherapy and cytokine therapy are preferred to prevent the recurrences. Chemotherapy application is still controversial. All these adjuvant treatment procedures may be adopted in case of small tumours or for surgical support\(^{(10,14)}\). In cases of scalp involvements, cranium and dura are the natural barriers for cerebral invasion. The prognosis of the young patients is better. The metastasis detection in the first diagnosis demonstrates the poor prognosis. 5 year survival rates are less than 10\% – 30\%. Local invasion, high recurrence rate, malignant biological tendency make the early diagnosis and treatment of these rare tumours important\^{(20,23)}.

CONCLUSION

Angiosarcoma is an extremely aggressive tumour which involves the scalp and has poor prognosis. Early diagnosis and radical treatment are substantial. Early diagnosis may be delayed due to its low prevalence in the older population. Large surgery is of significance to prevent the resection recurrence when the diagnosis is established. If the tumour is less than 5 cm in size, prognosis is better. Radiotherapy is necessary after large surgical resection. Due to the high tendency of recurrence and distant metastasis, a close follow up is required.

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